A lifetime of specialist care

Royal Brompton & Harefield NHS
NHS Foundation Trust

Adult Congenital Heart Disease
Echocardiographic Evaluation

Dr. Wei Li

Royal Brompton Hospital and National Heart Lung Institute,
Imperial College, London, UK

A lifetime of specialist care

Introduction

Adults with CHD represent a unique patient population

- Great variation of anatomic lesions
- Large majority had undergone previous palliative or “corrective” surgery
- Ventricles are exposed to abnormal physiological conditions which may dominate the clinical picture, later on in life
- Continuous evolution of surgical techniques and catheter interventions over the past 60 years have created a special patient population.

Sequential Segmental Analysis

1. Position of the heart and direction of apex

   Levocardia, Dextrocardia, Mesocardia

2. Arrangement of atrial chambers

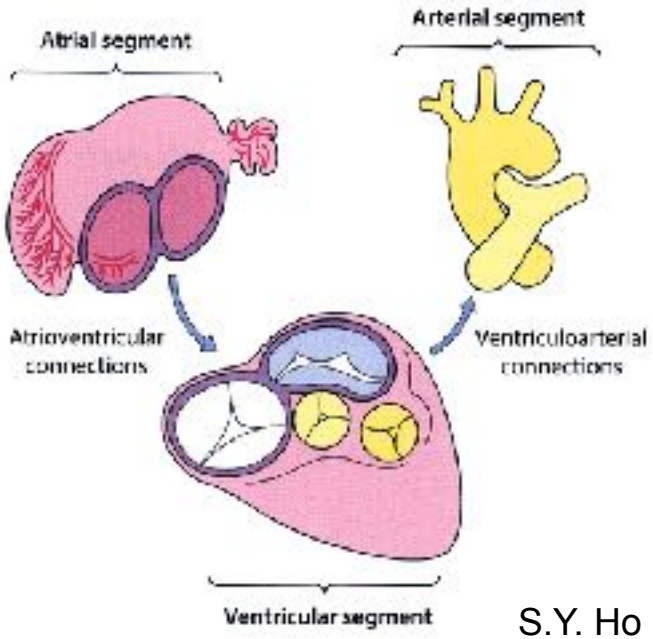
   situs solitus, situs inversus, situs ambig. (isomerism)

3. Atrioventricular junctions, V. morphology

   AV concordance, AV discordance, Double inlet, absent connection

4. Ventriculo-arterial relationship

   VA concordance, discordance, Double outlet, Solitary Arterial trunk



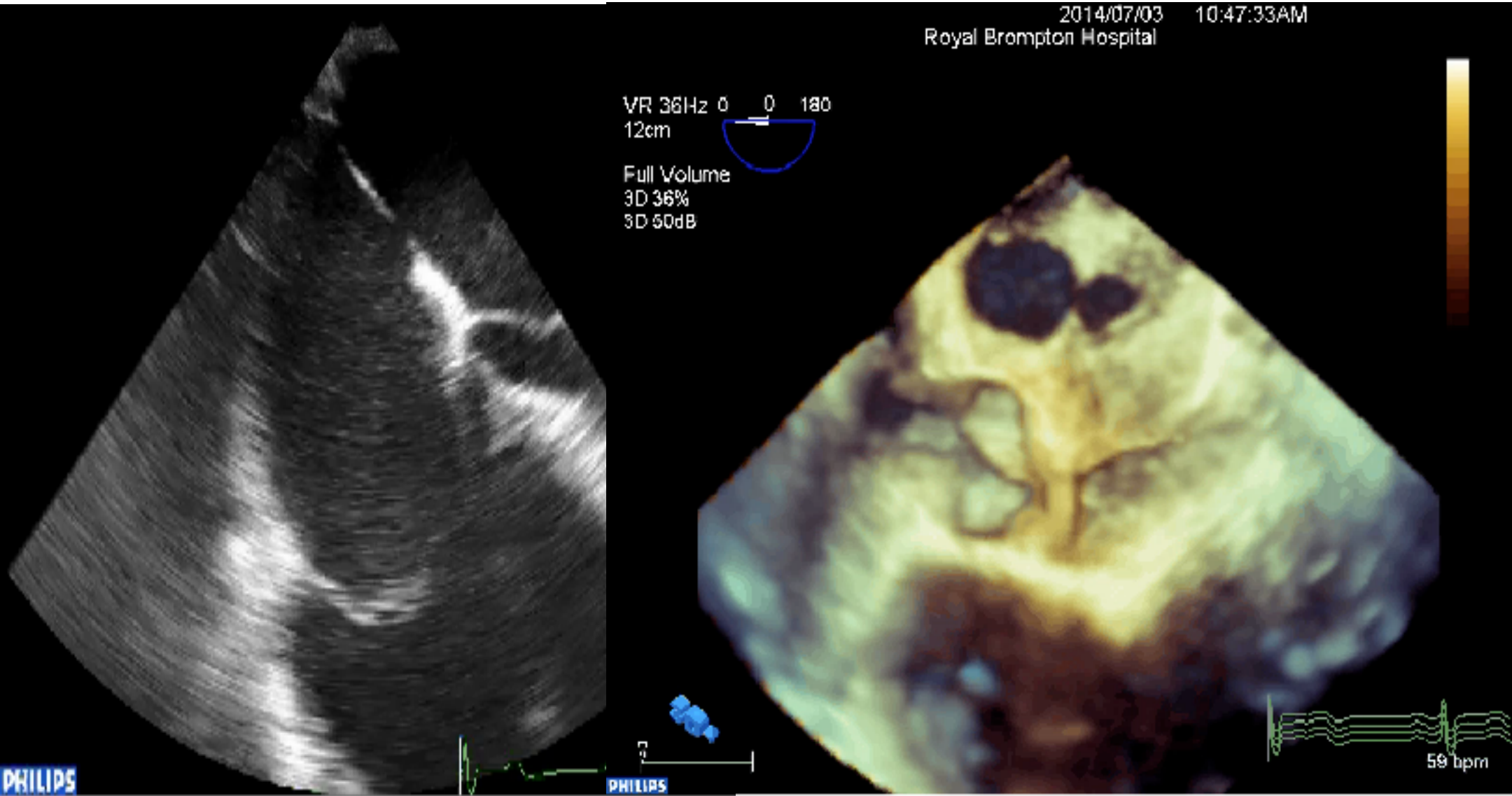
S.Y. Ho

Simple Congenital Heart Disease

- **Left to right shunt lesions**
  1. **Atrial Septal Defect**
  2. **Ventricular Septal Defect**
  3. **Atrioventricular Septal Defect**
- **Left ventricular outflow tract lesions**
- **Diseases of aorta**
- **Diseases of tricuspid valve**

Abnormal intra-cardiac communication

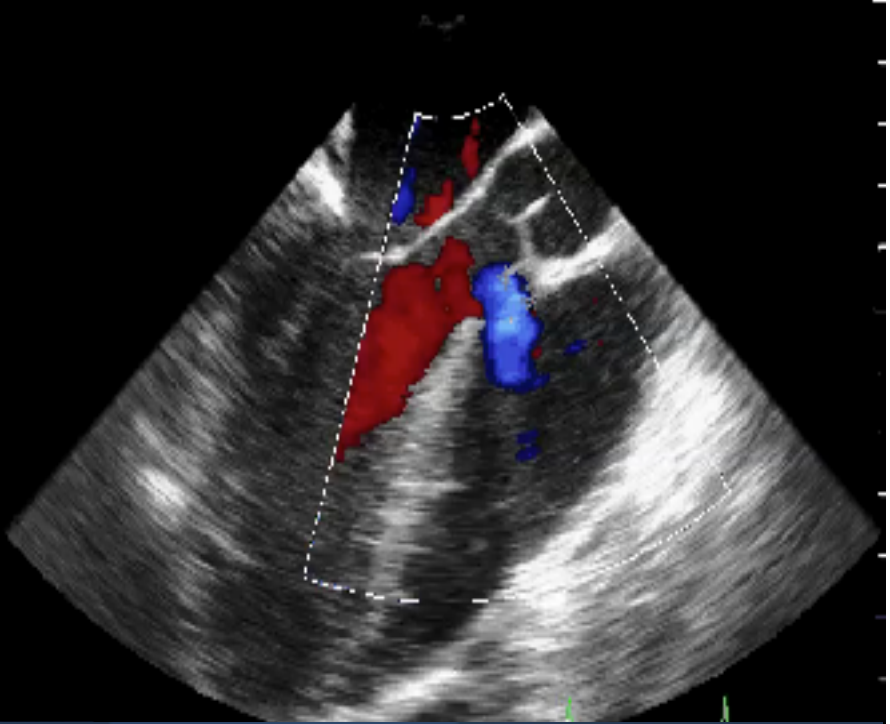
ASD – multiple defects



Ventricular Septal Defect in the Adult Population

- Small (restrictive) VSD
- Large VSD with Eisenmenger physiology
- VSD with significant L-R shunt
  - LV volume overload
  - +/-pulmonary hypertension
- Repaired VSD +/-residual shunt

Small “Restrictive” VSD



Vmax across VSD > 4 m/s ?

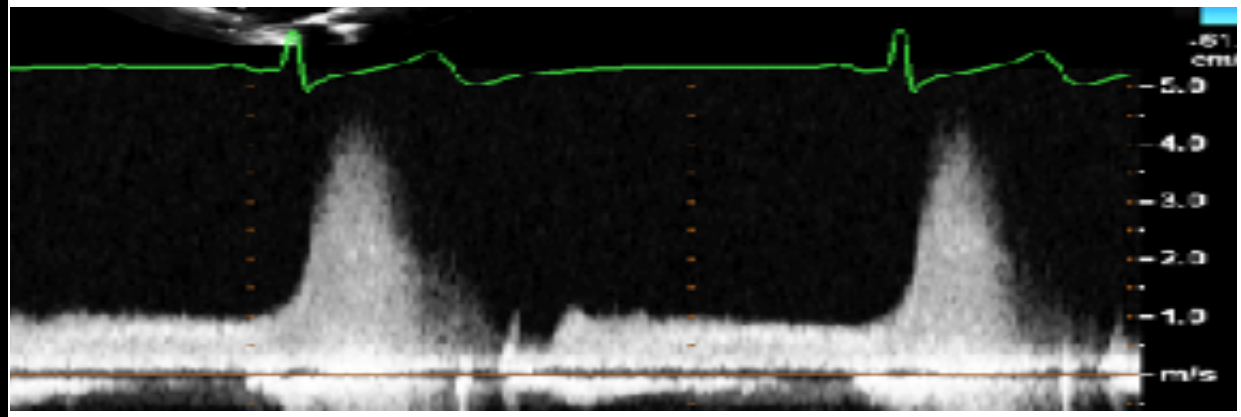
L-R shunt may ↑ with time

Double-chambered RV (13%)

LV disease (10%)

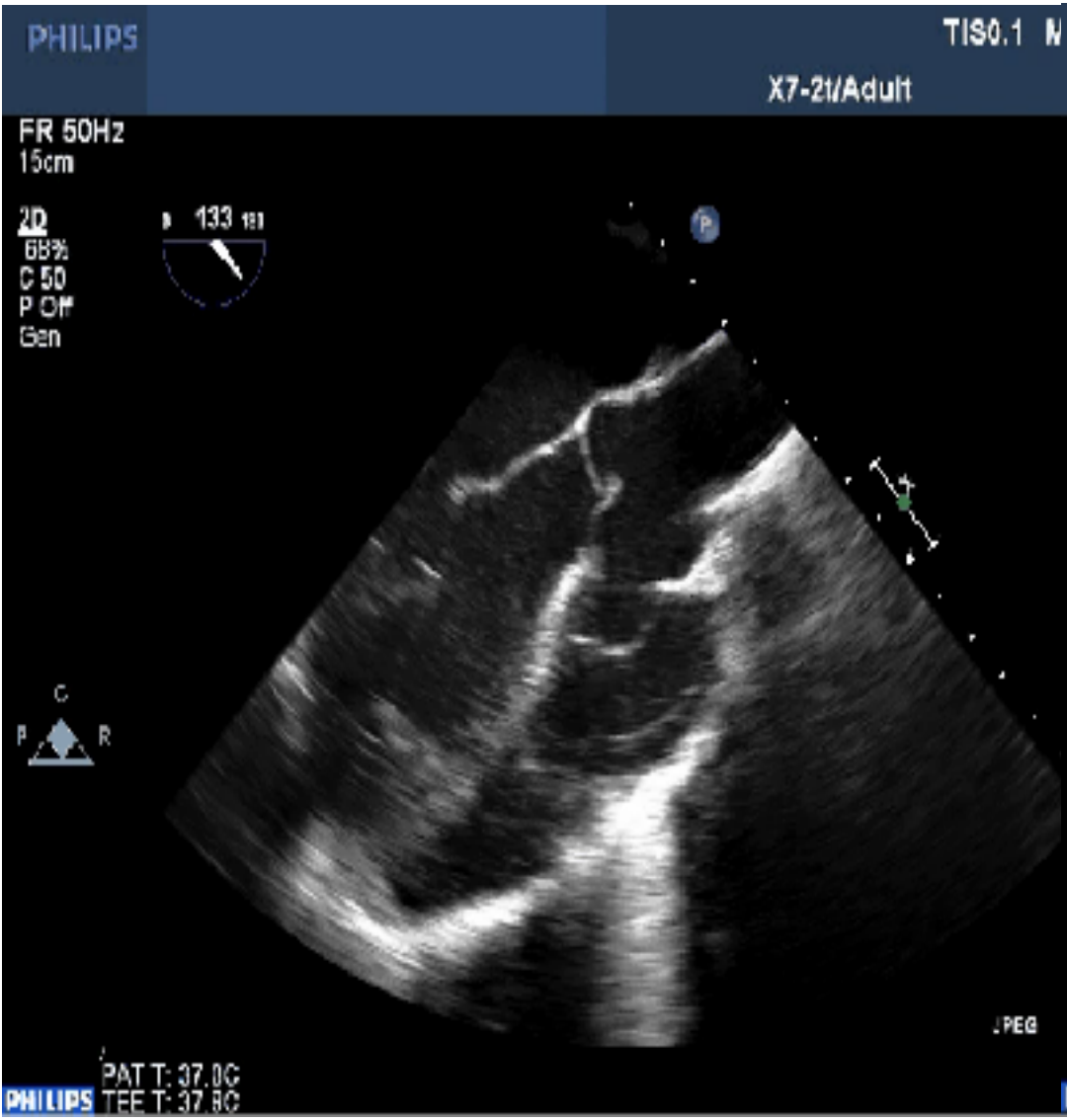
Risk of endocarditis is high (10%)

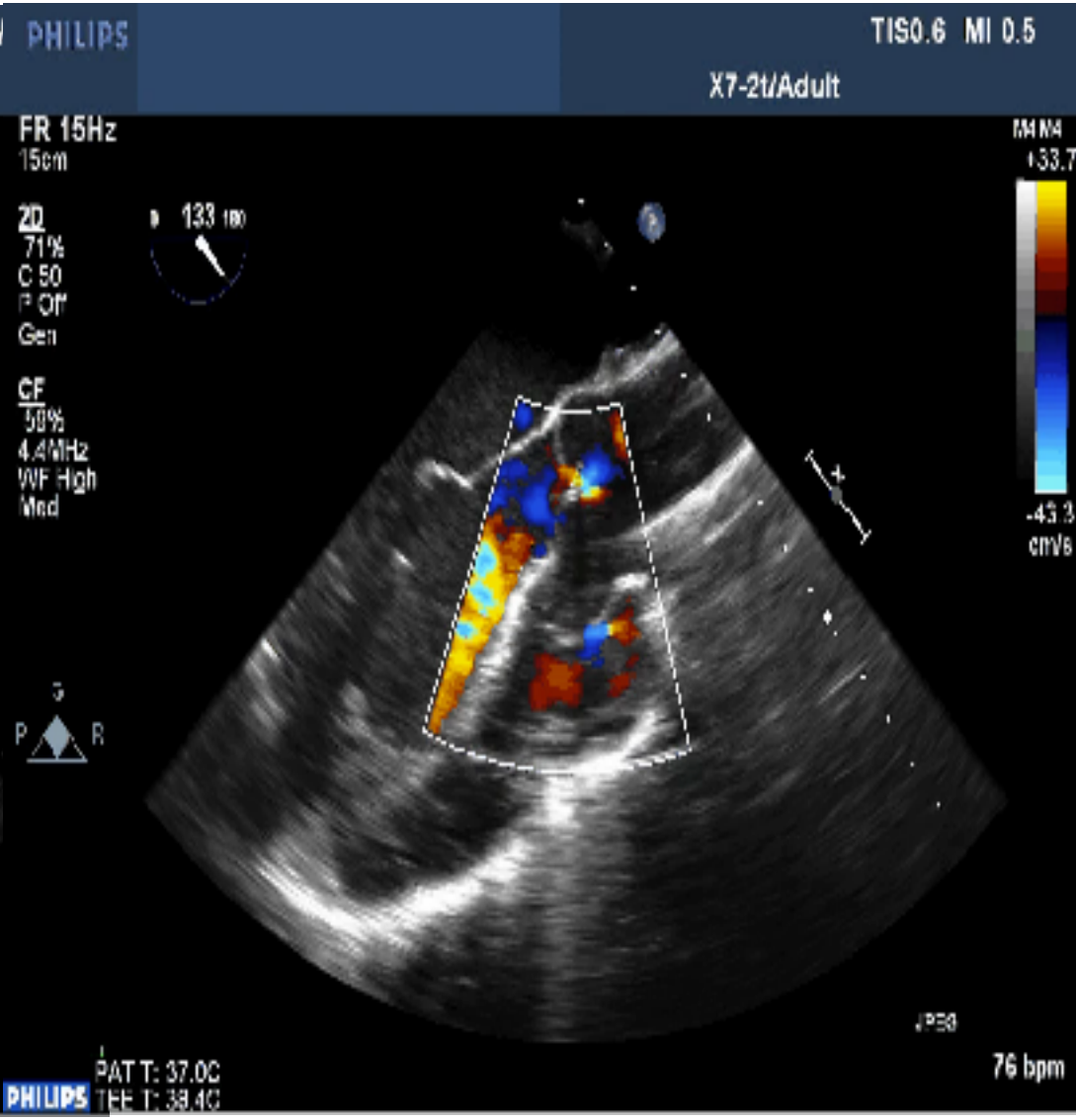
Sig. Aortic regurgitation (3%)

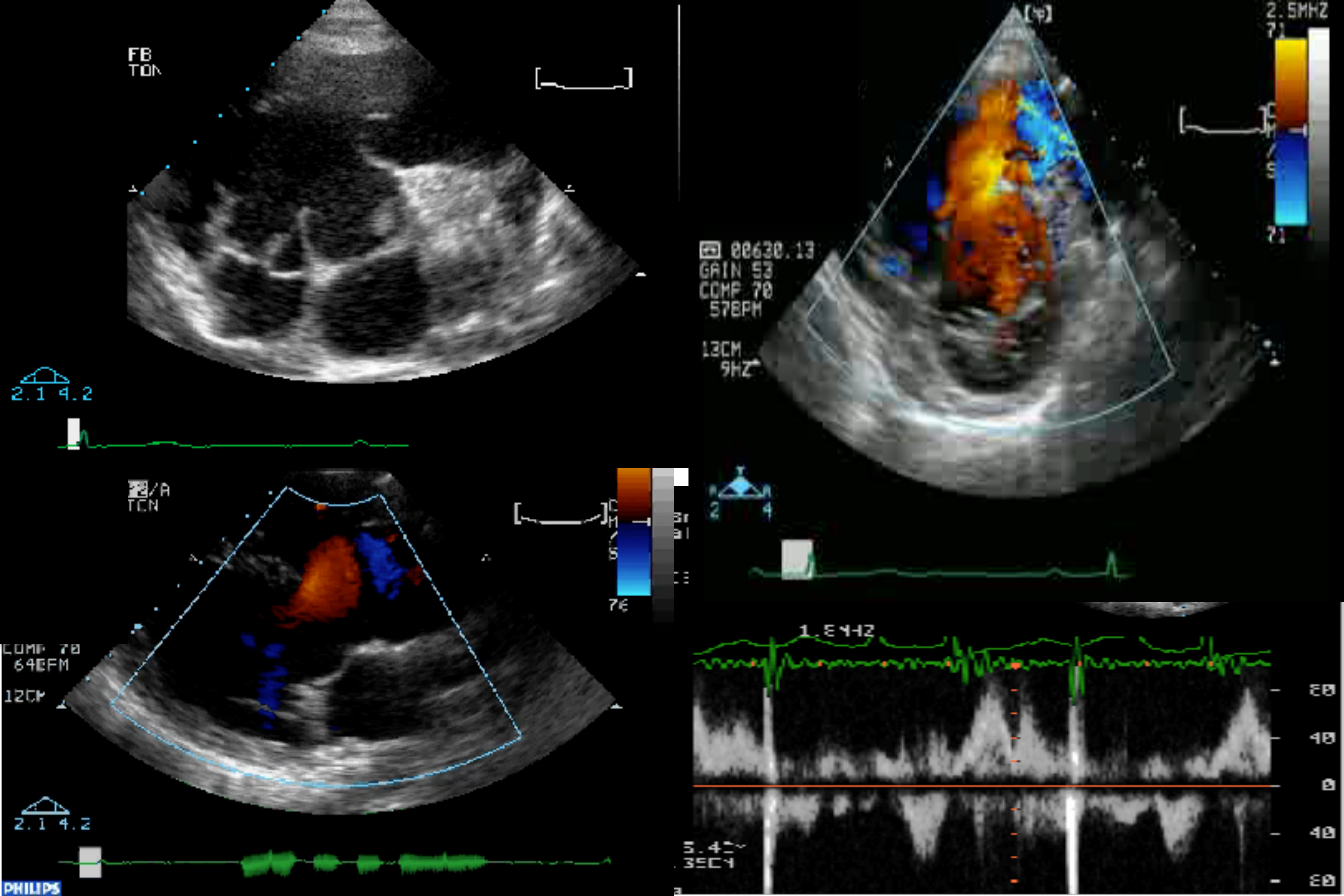


Karonis et al, IJC 2016, 102-6

Perimembranous VSD and AR






FB
2.1 4.2
2.5MHZ
GAIN 53
COMP 70
57BPM
12CM
9HZ
COMP 70
64BPM
12CM
2.1 4.2
80
40
0
40
80
PHILIPS

Eisenmenger Syndrome

All lesions with systemic-to-pulmonary shunts due to large defects leading to a severe increase in PVR, and initial L-R shunt reverses direction.

- Prognosis is good (much better than primary pulmonary hypertension)
- Closure of defects is strongly contraindicated
- Pregnancy is contraindicated
- Risks of GA are very high
- No place for venesection

Ebstein anomaly - Definition

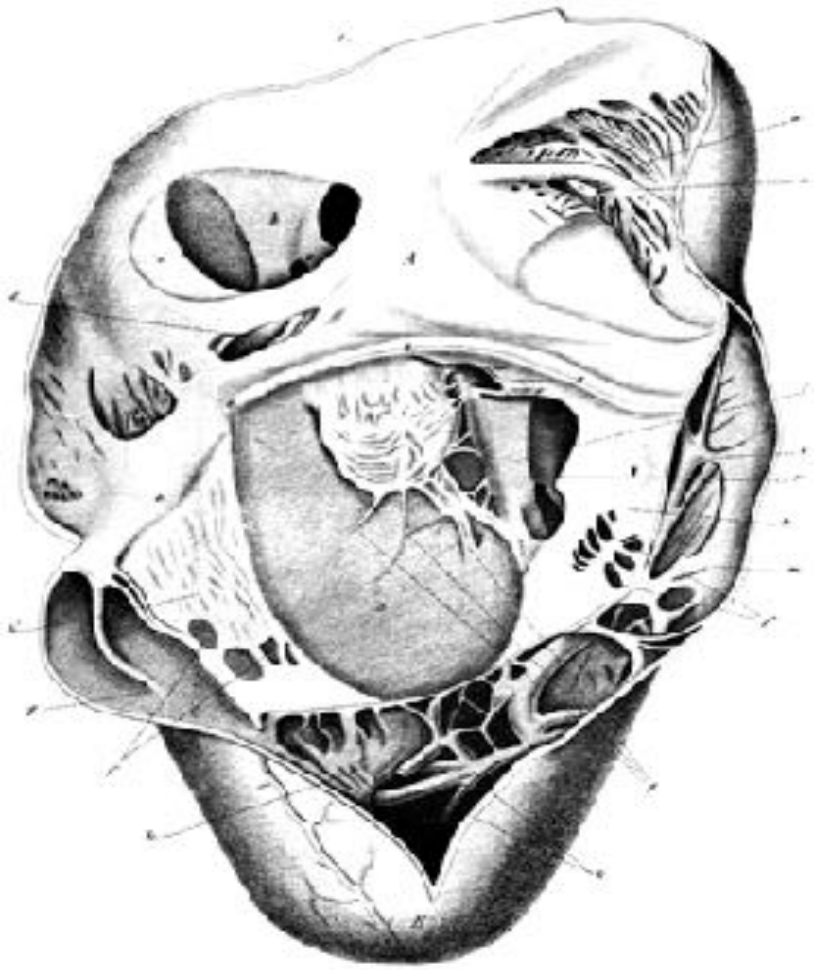


Figure from Ebstein's original case report

- Apical displacement of the septal and posterior leaflets of the TV into the RV. Leaflets often dysplastic, thickened-rolled, short chordae and underdeveloped papillary muscle
- Anterior leaflets are elongated and redundant with abnormal chordae attachment.
- "Atrialization" of the basal portion of the RV.

Attenhofer Jost C H et al. Circulation 2007;115:277-285

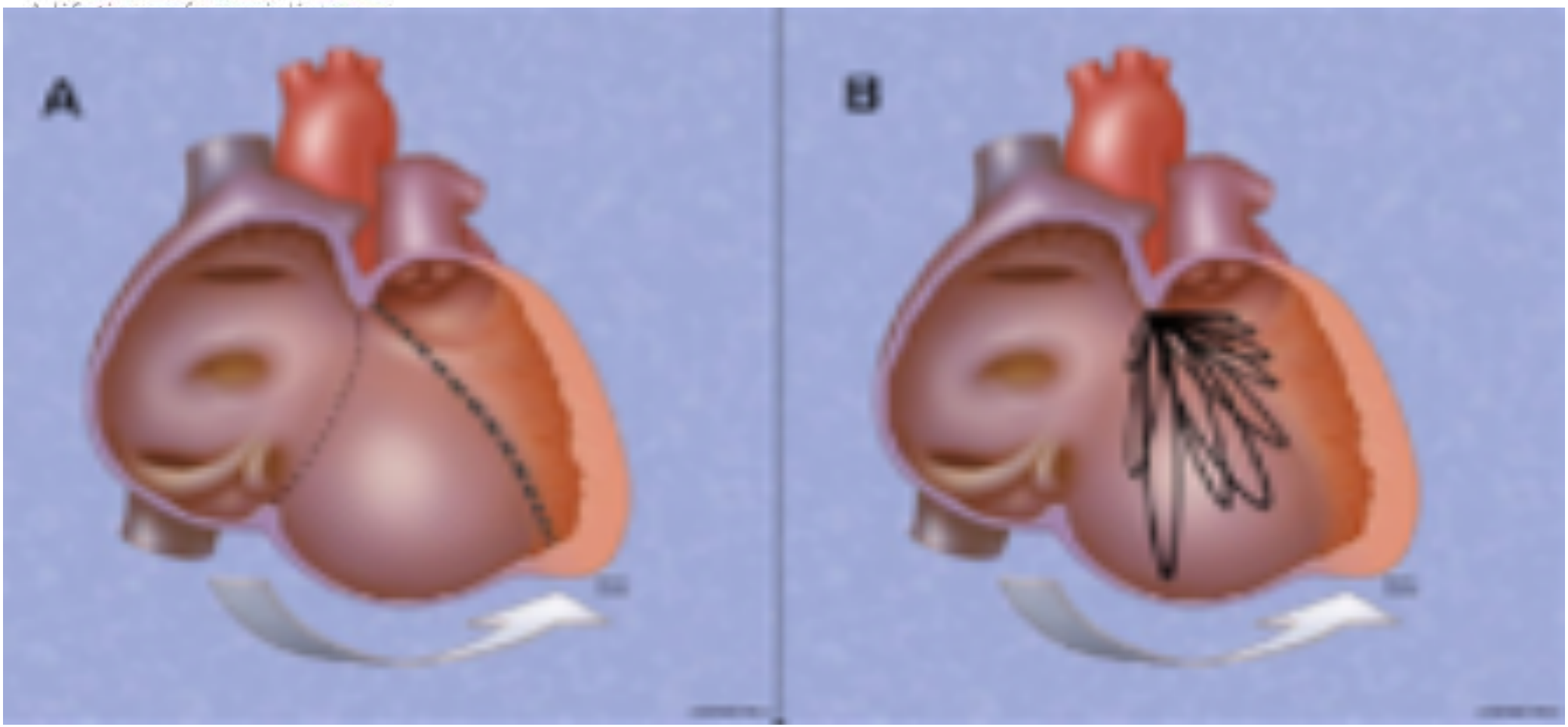
Echocardiographic Diagnosis

- Displacement of the septal TV leaflet (best seen from Apical four camber view)

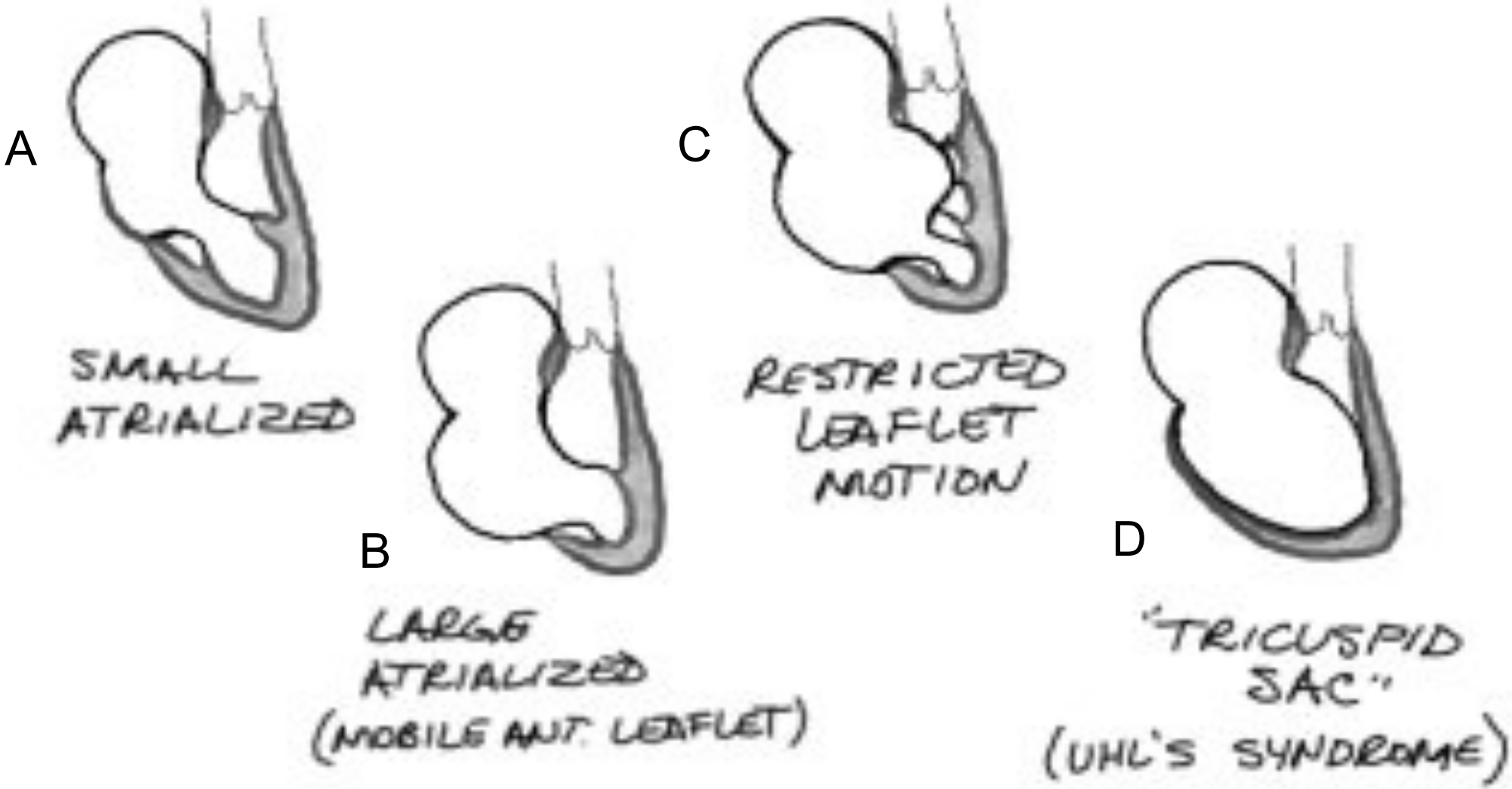
  > 8mm/M^2 BSA

  or > 20 mm

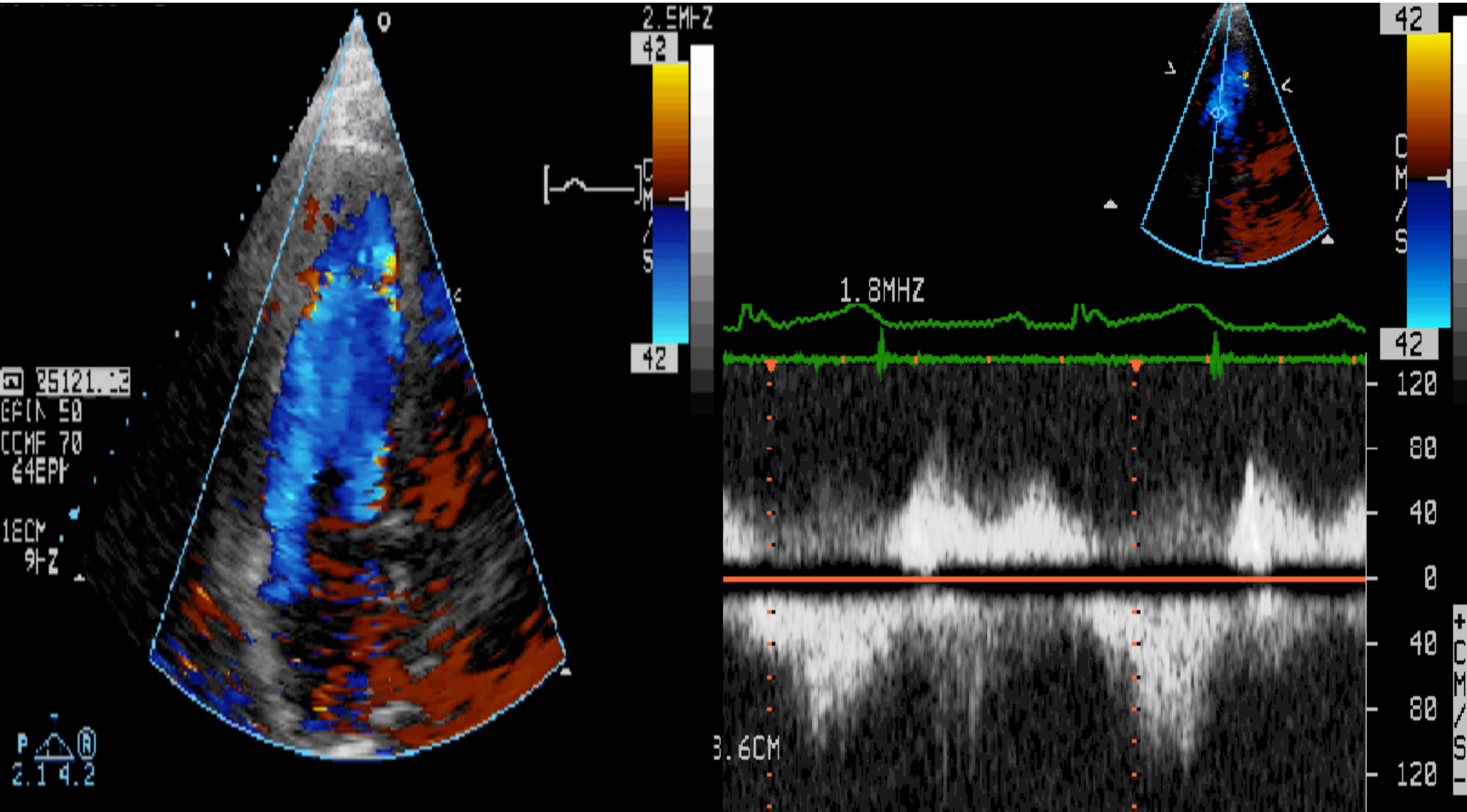
  is indicative of Ebstein anomaly.
- Anterior leaflet: elongation, redundant and/or sail-like.


A
B

CARPENTIER'S CLASSIFICATION



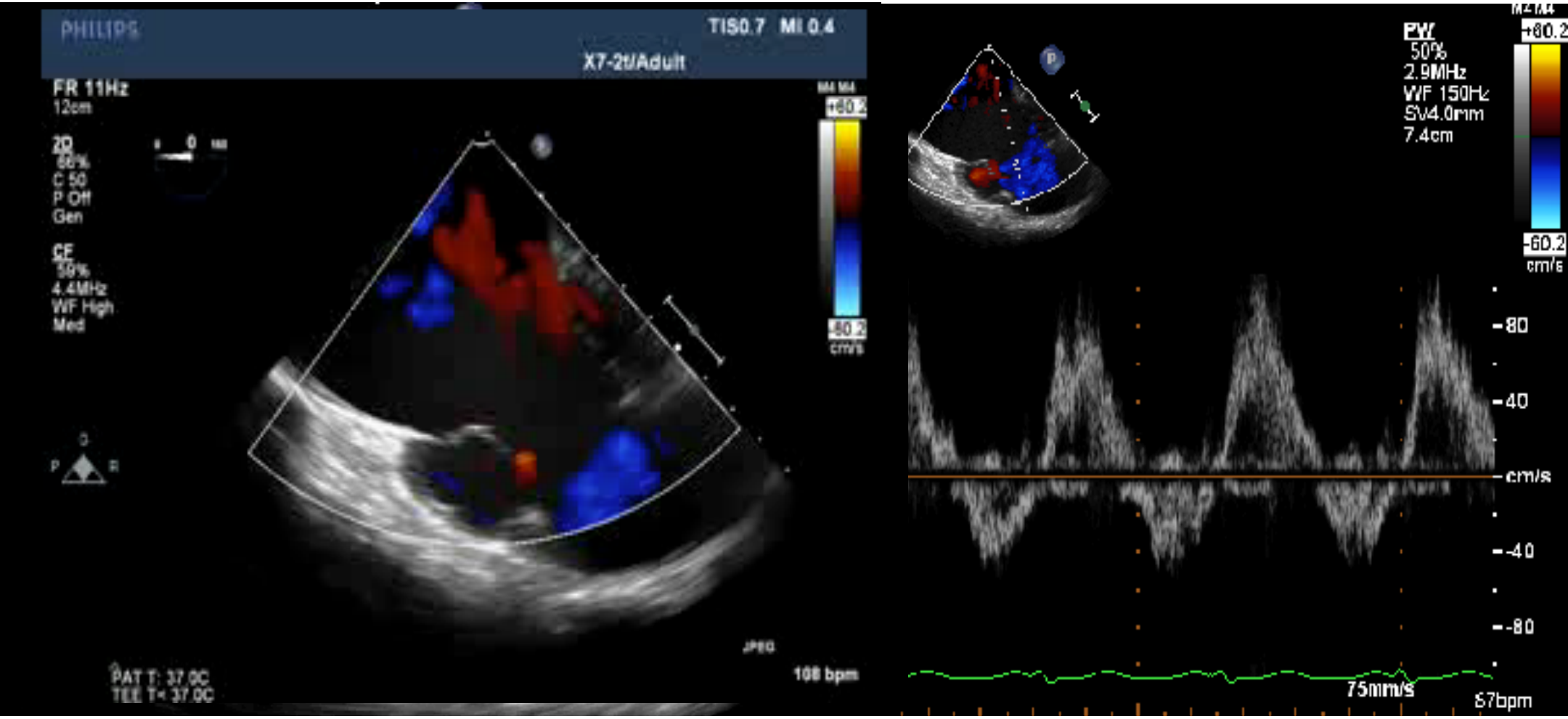
Tricuspid Valve Regurgitation



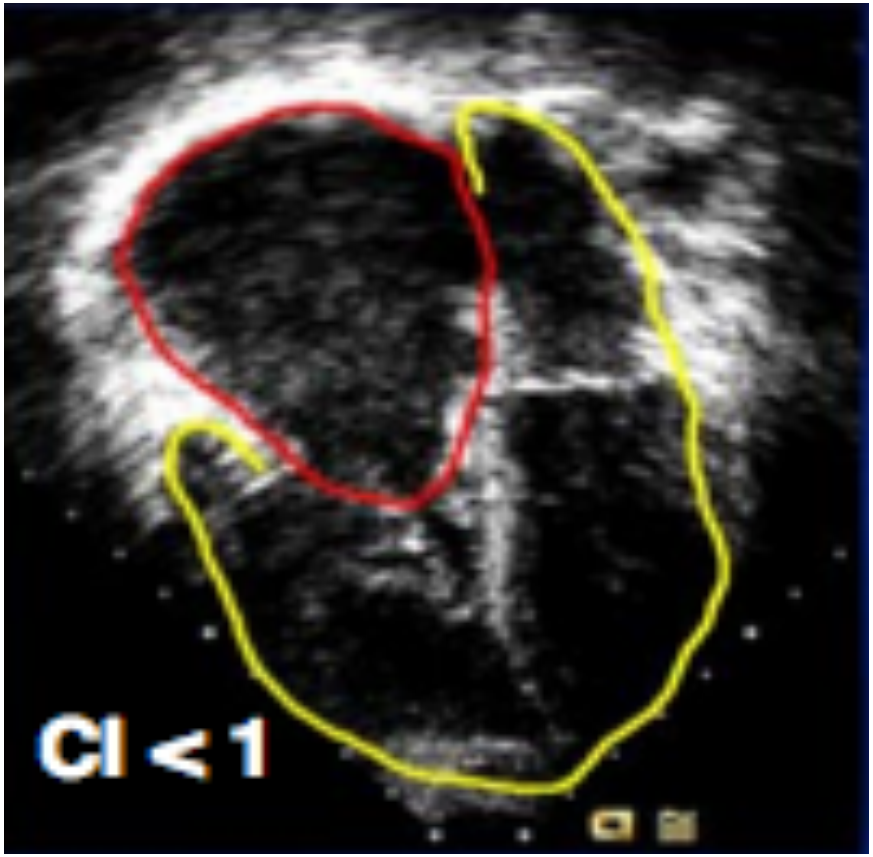
A lifetime of specialist care

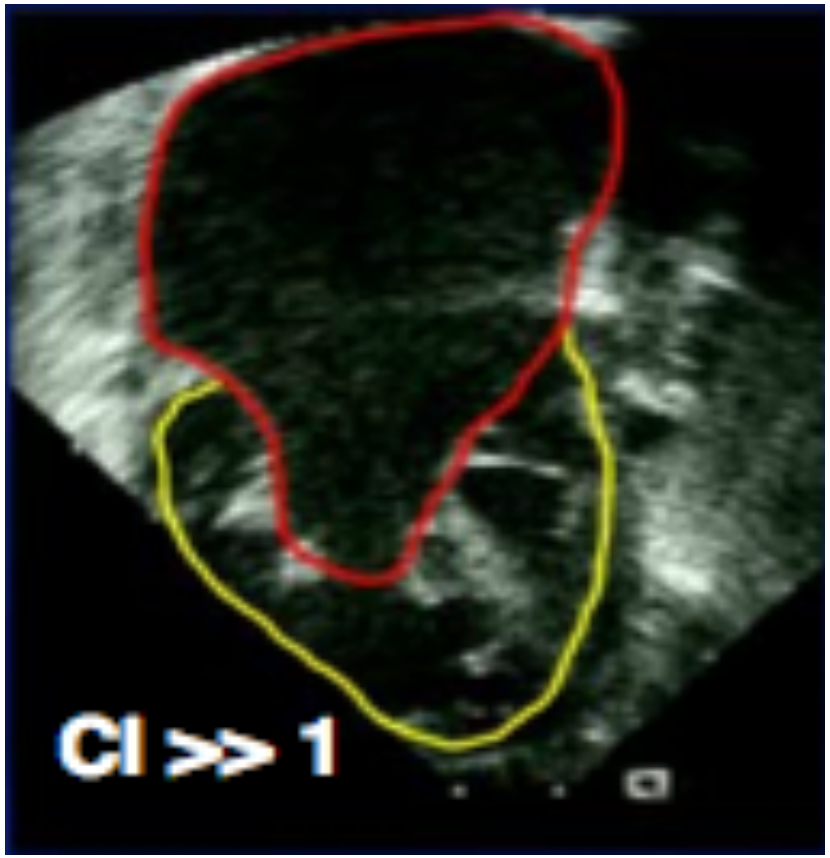
Royal Brompton & Harefield NHS
NHS Foundation Trust

Tricuspid valve regurgitation



Celermajer Index in Ebstein Anomaly

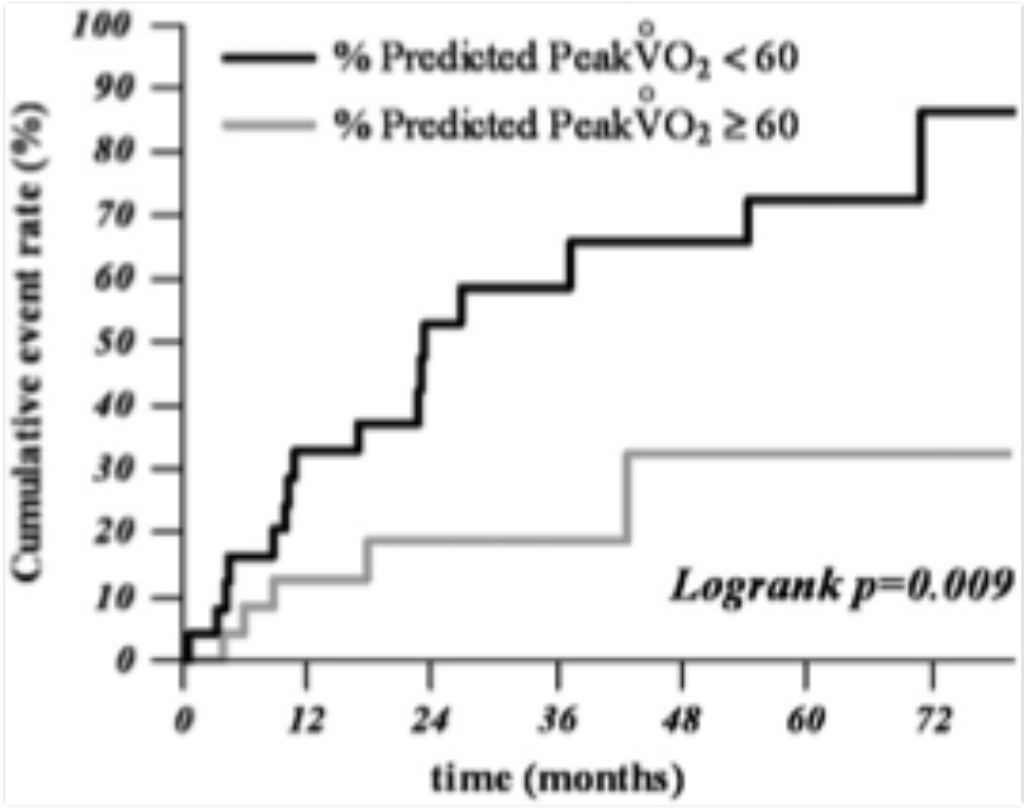




(RA + aRV)/(RV+LA+LV)

Celermajer D et al. J Am Coll Cardiol 1992: 19:1041-6

Ebstein anomaly of tricuspid valve



Reduced peak VO2 related to

Glasgow Score

Cyanosis (ASD /PFO)

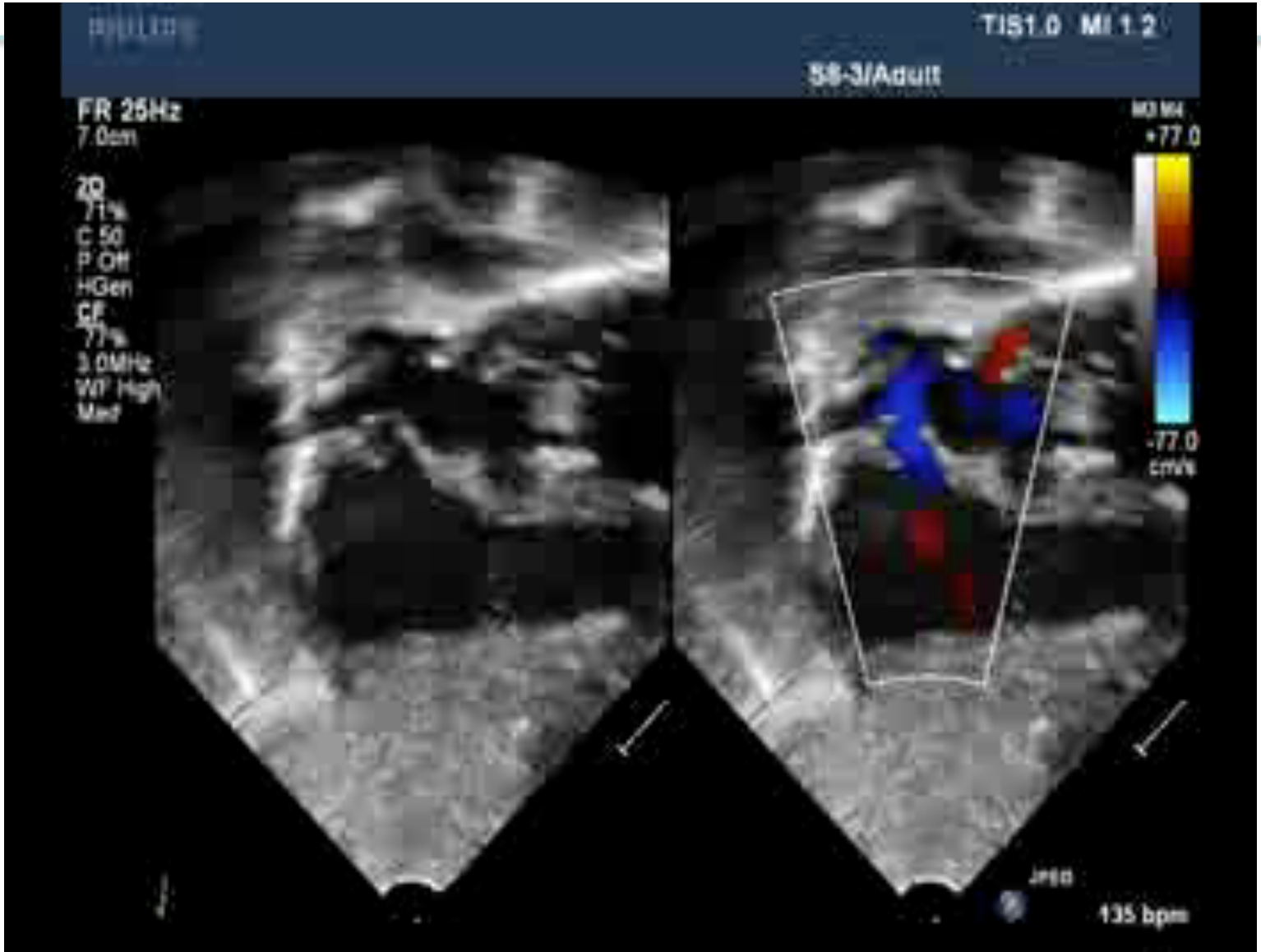
Cardiomegaly

Radojevic et al, IJC Vol 163 (2013): 305-308

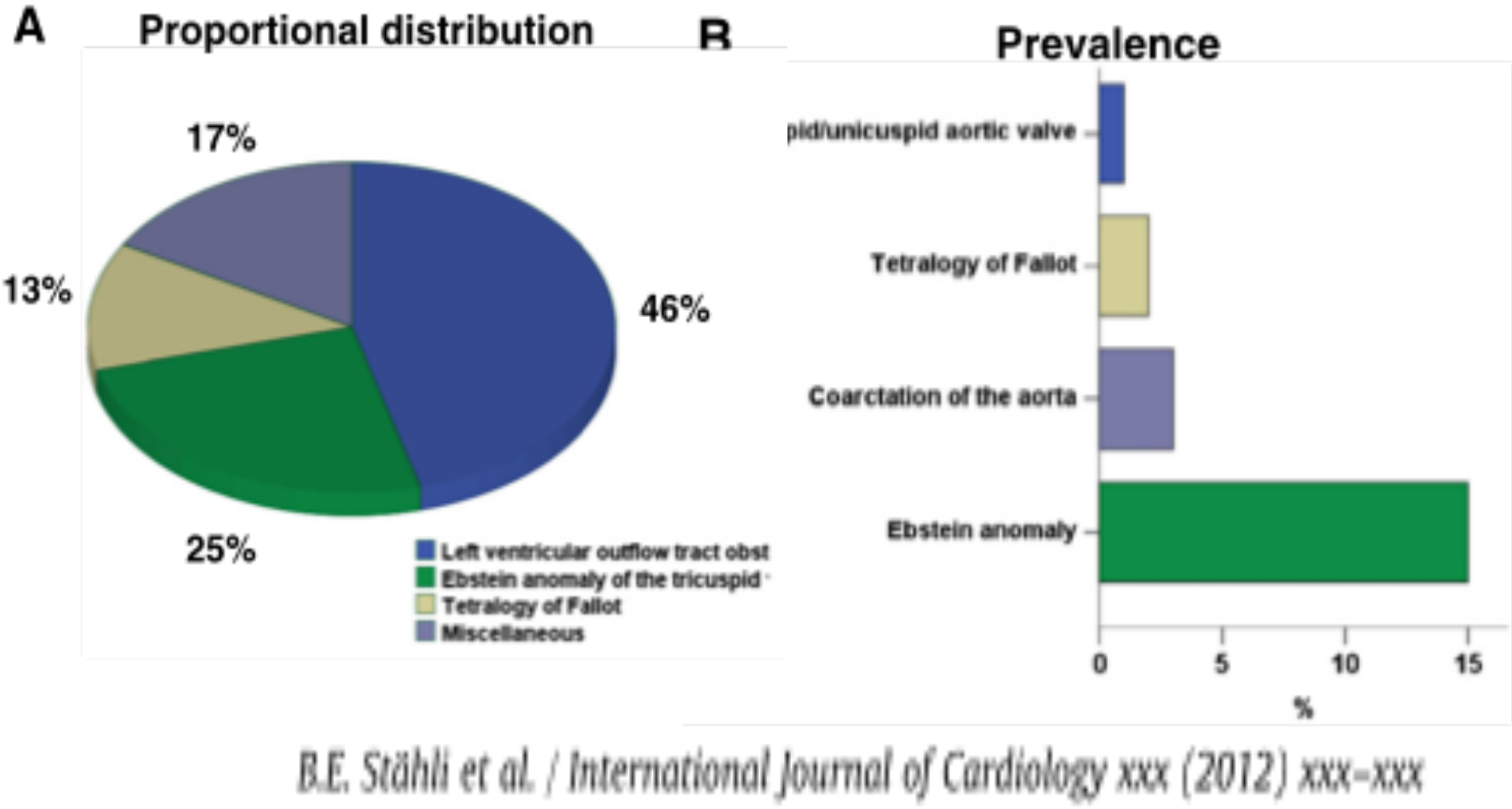
Associated Lesions

- PFO/ASD
  - Left to right shunt – Additional RV volume overload
  - Right to left shunt – cause of cyanosis
- Pulmonary stenosis
- LV non-compaction
- Bicuspid aortic valve
- LVOTO

ASD/PFO with R-L shunt



Left Ventricular Non-Compassion in CHD



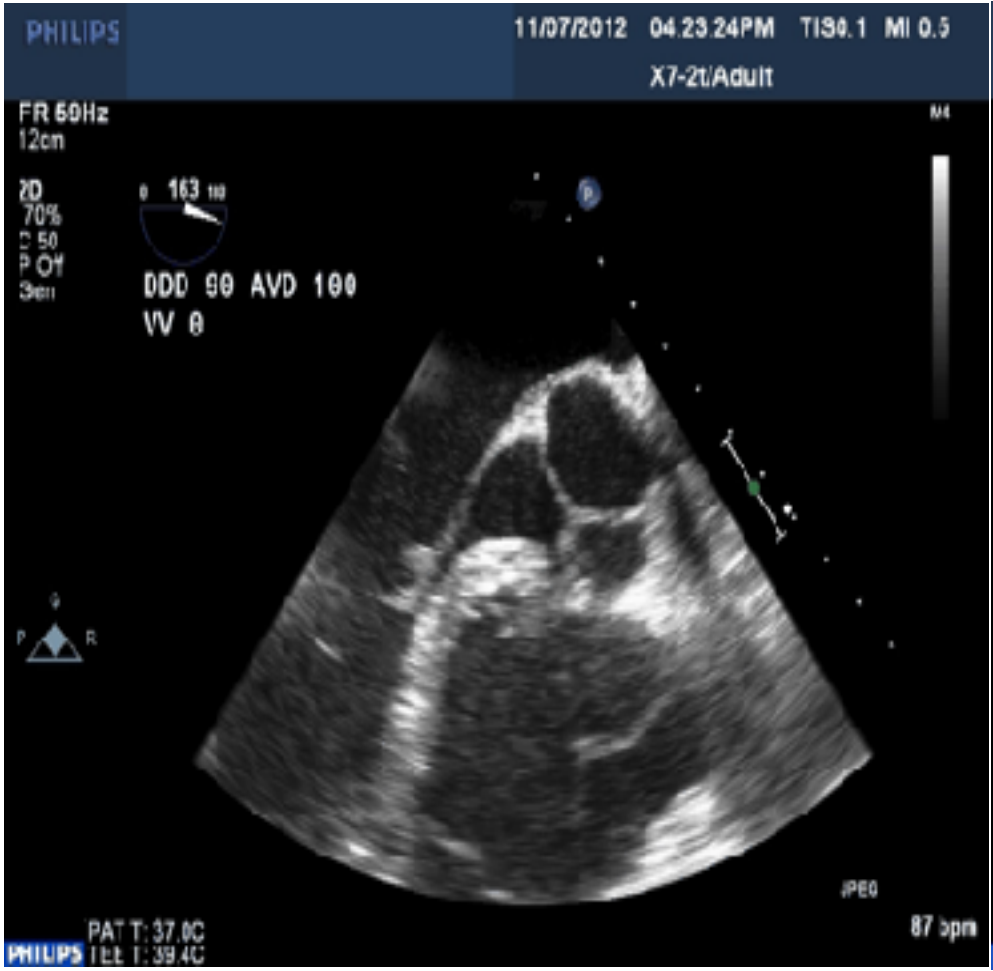
B.E. Stähli et al. / International Journal of Cardiology xxx (2012) xxx–xxx

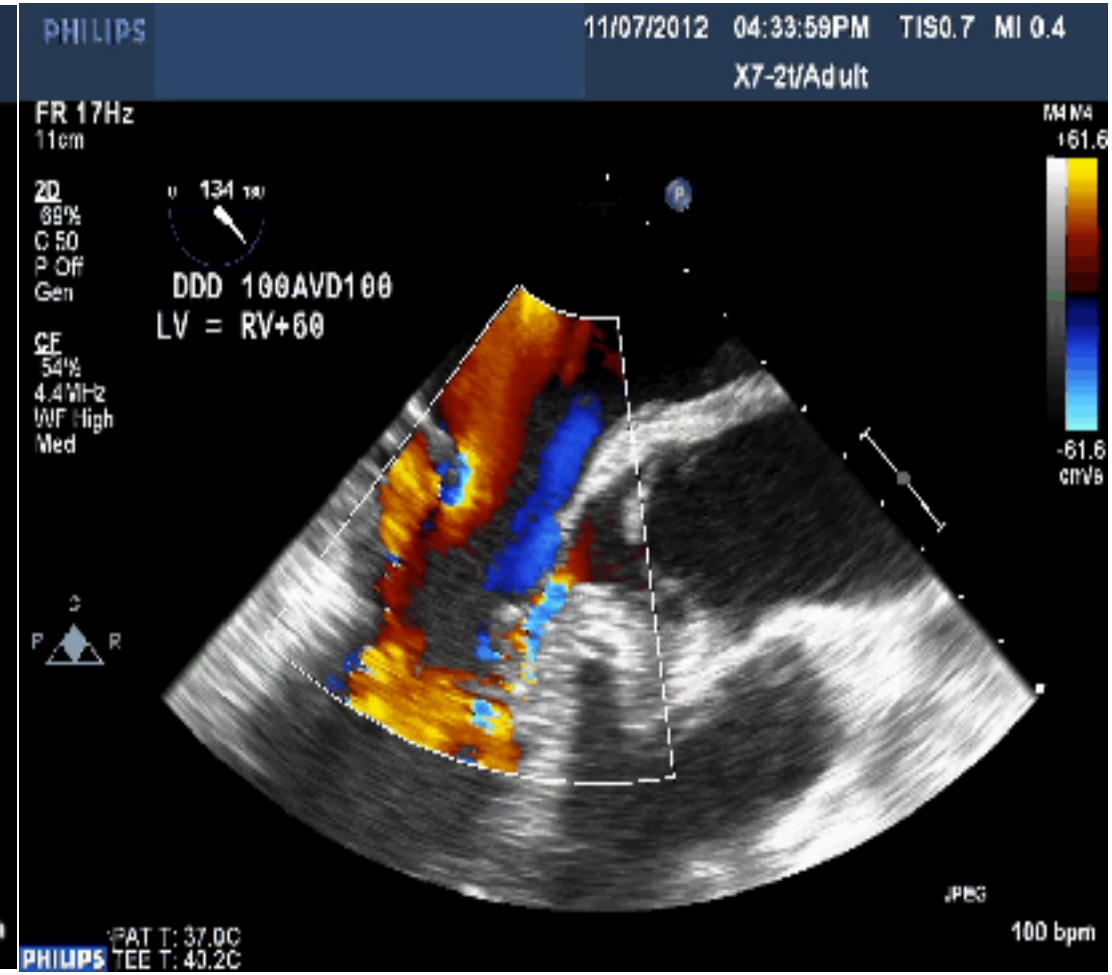
Ebstein anomaly with non-compaction

A lifetime of specialist care

Royal Brompton & Harefield NHS
NHS Foundation Trust

Ebstein Anomaly - LVOTO





Complex Congenital Heart Disease

1. Repaired Tetrology of Fallot
2. Transposition of great arteries
    - Post atrial switch
    - Post arterial switch
    - Rastelli repair
3. Univentricular heart

Assessing the Adults with Repaired TOF

- Quantify pulmonary regurgitation
- Image RVOT, PA branches
- Bi-ventricular size and function
- Estimate RV systolic pressure
- Residual VSD, BT shunts or MAPCAs
- Assess aortic root size, AR ?

Quantification of pulmonary regurgitation

1. Diastolic flow reversal from MPA and branches
2. PR jet width
3. PR Deceleration time
4. PR index
5. RPA pulsatility (systolic to diastolic diameter ratio)

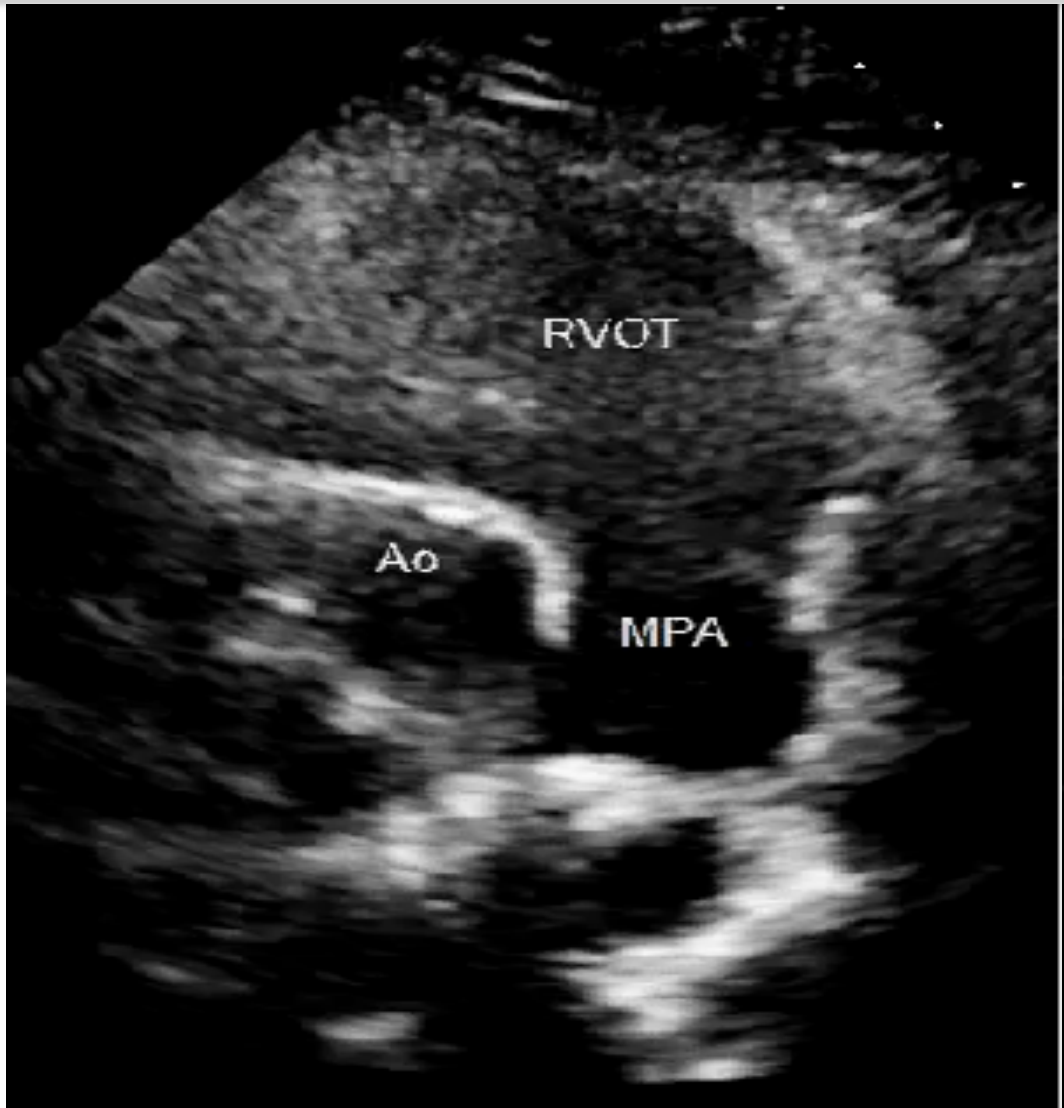
Parasternal or subcostal

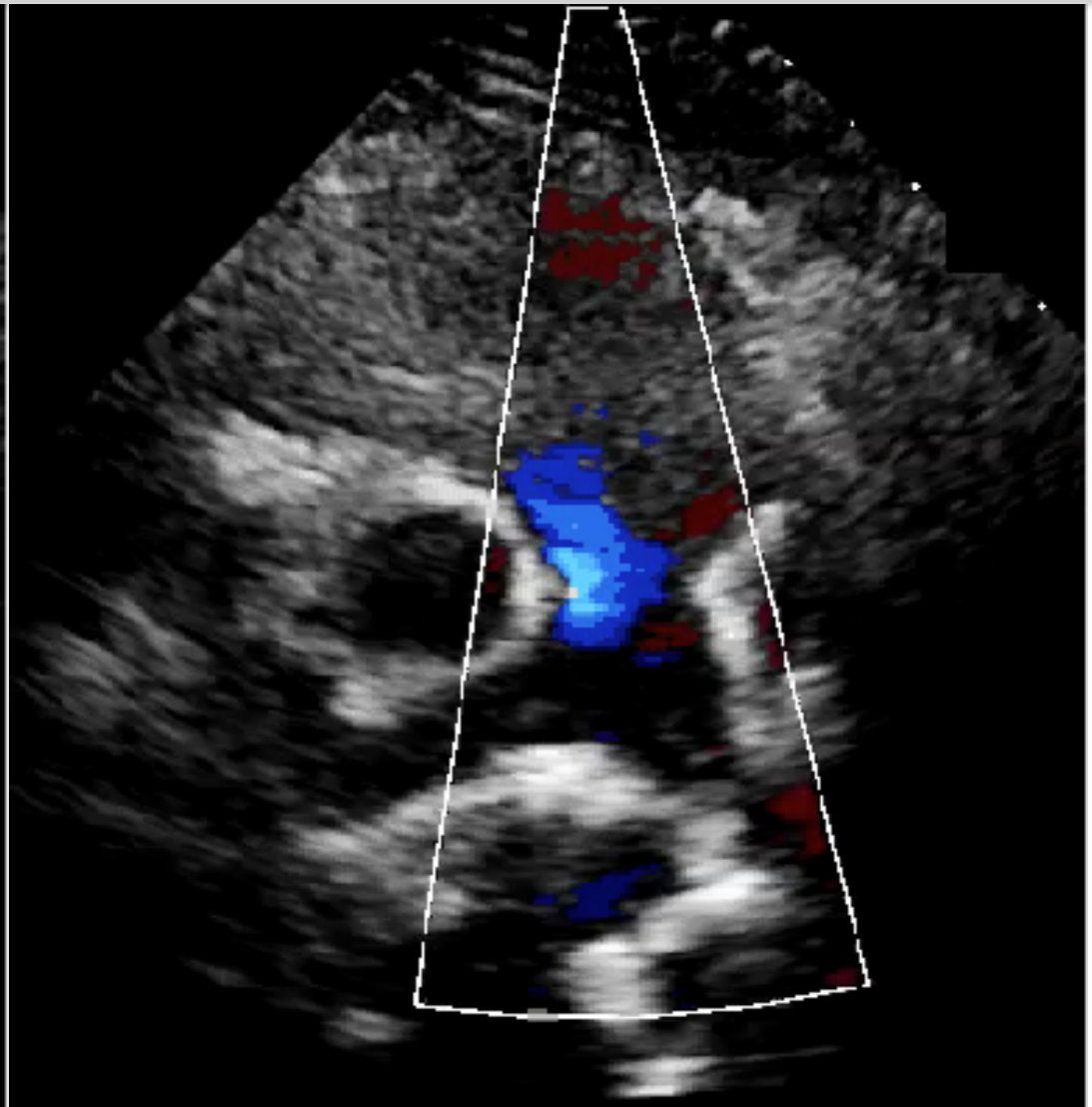
2D: Pulm. arteries / valve

Colour: turbulent /regurg.

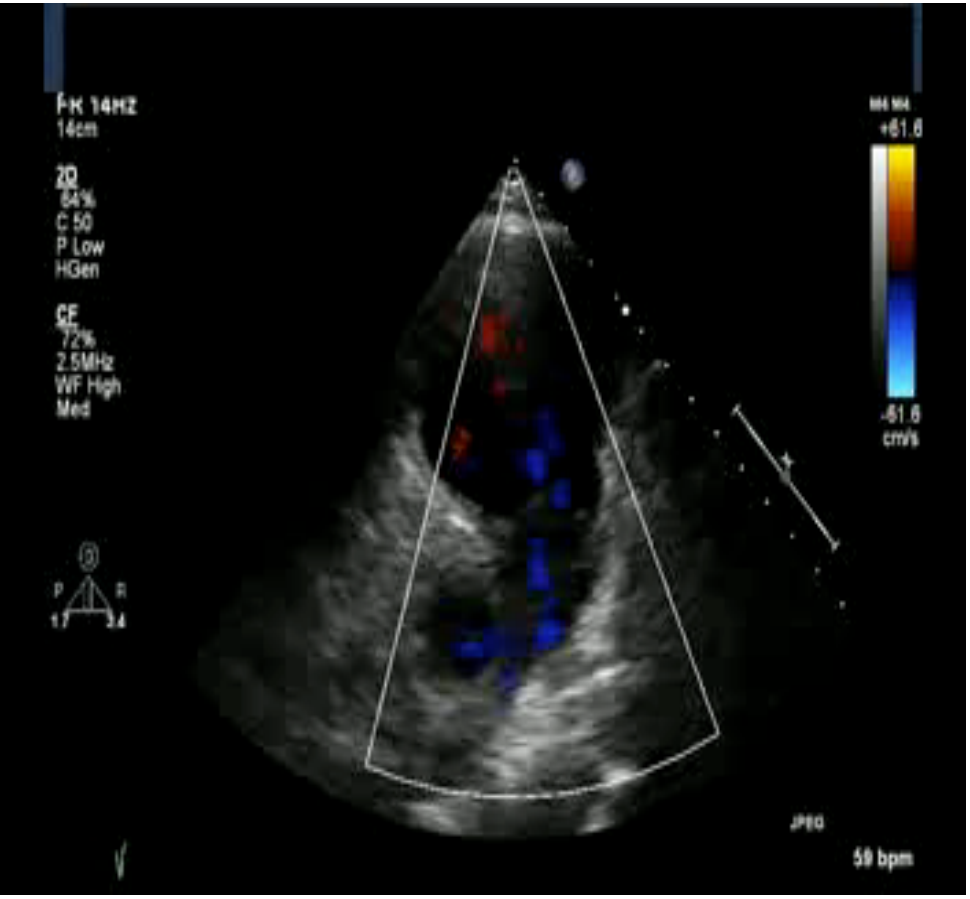
Doppler: flow velocity

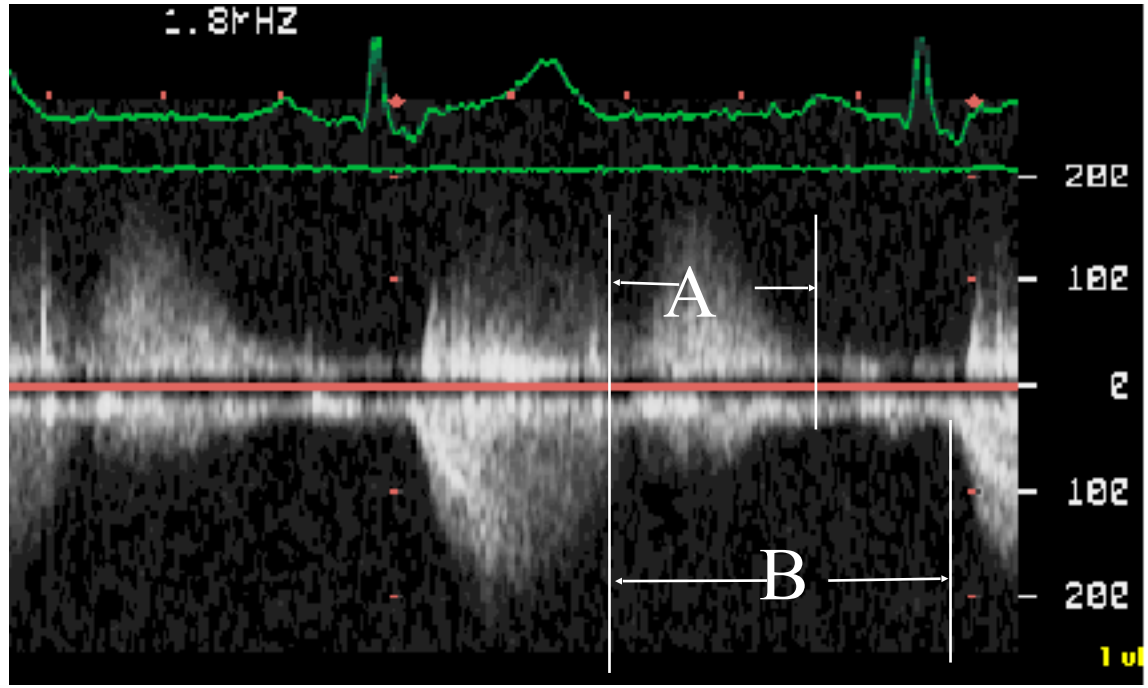
Pulmonary regurgitation





PR Quantification: Colour jet width and PR index

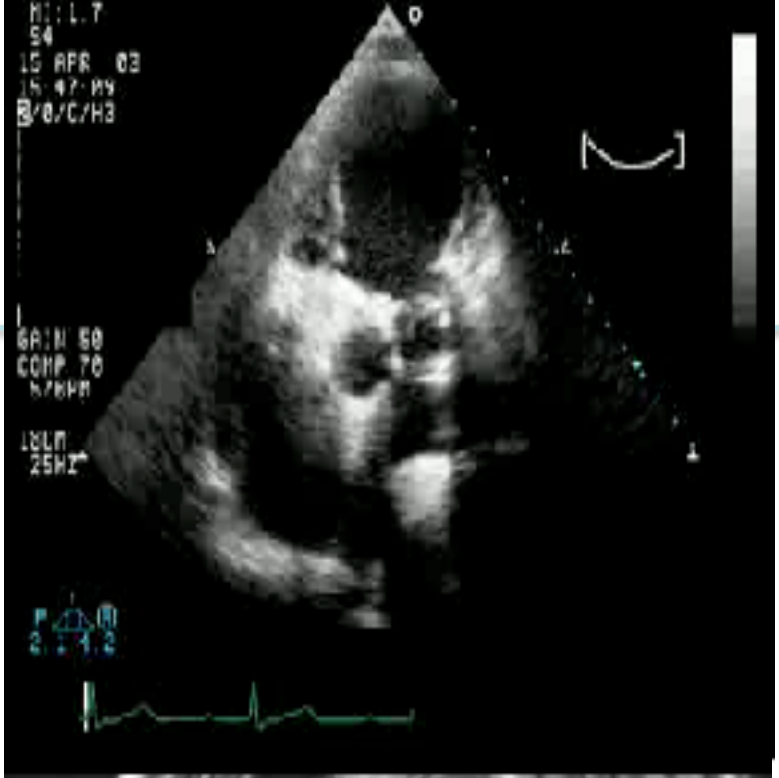


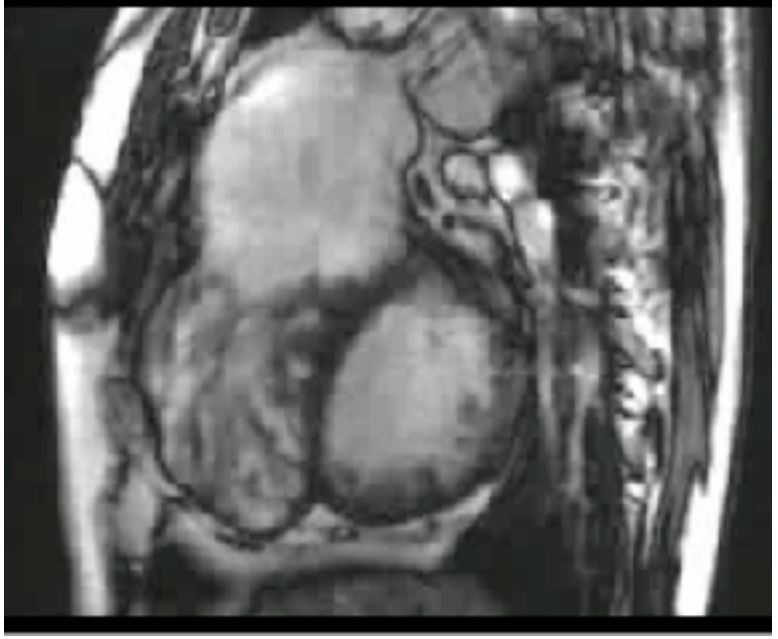


Significant PR:
Jet width>0.98

Severe PRi<0.7

Li et al: Am Heart J 2004:147:165-75





Repaired Fallot Assessment of RV

- Size and hypertrophy
- Outflow tract

  akinesis (~37%), aneurysmal (~18%)
- RV function

  Systolic function:

  RV FAC, TAPSE, S' Tei Index, 3D EF

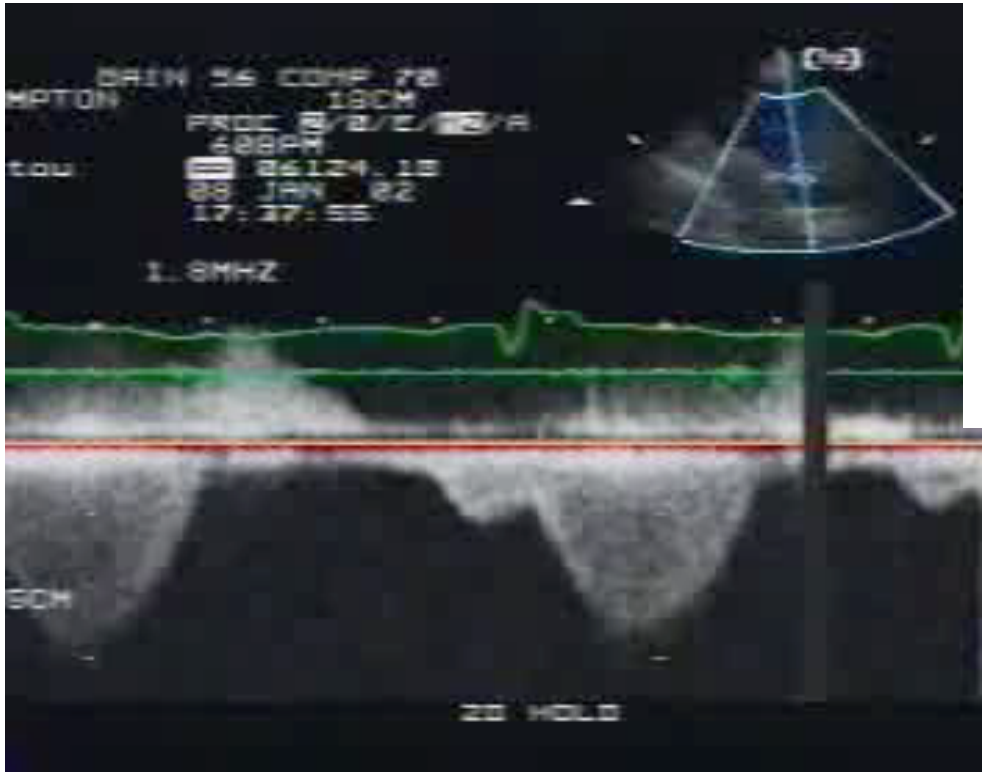
  Diastolic function:

  Diastolic forward flow in PA, Reversed flow in SVC

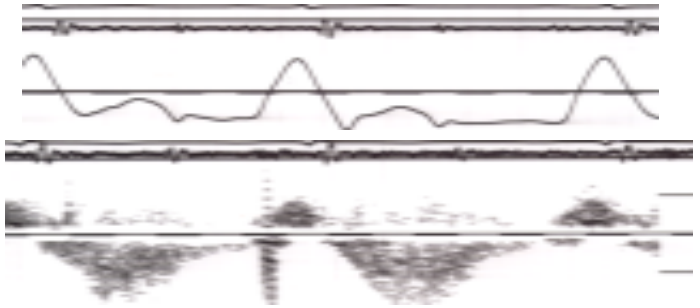
Bonello B et al: IJC 2013, 04,048

Repaired TOF - RV Function

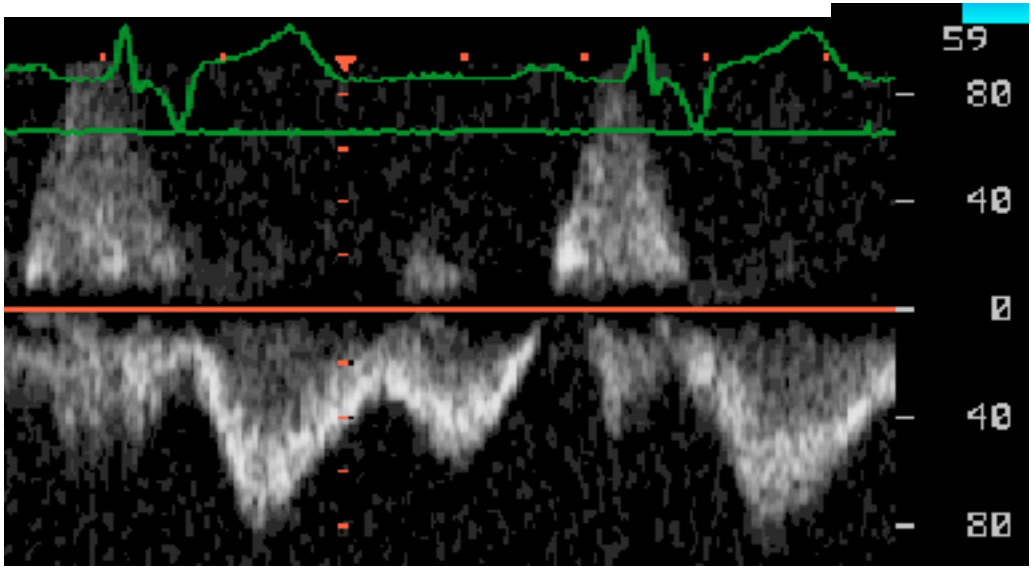
Restrictive RV Physiology



Pulmonary Doppler

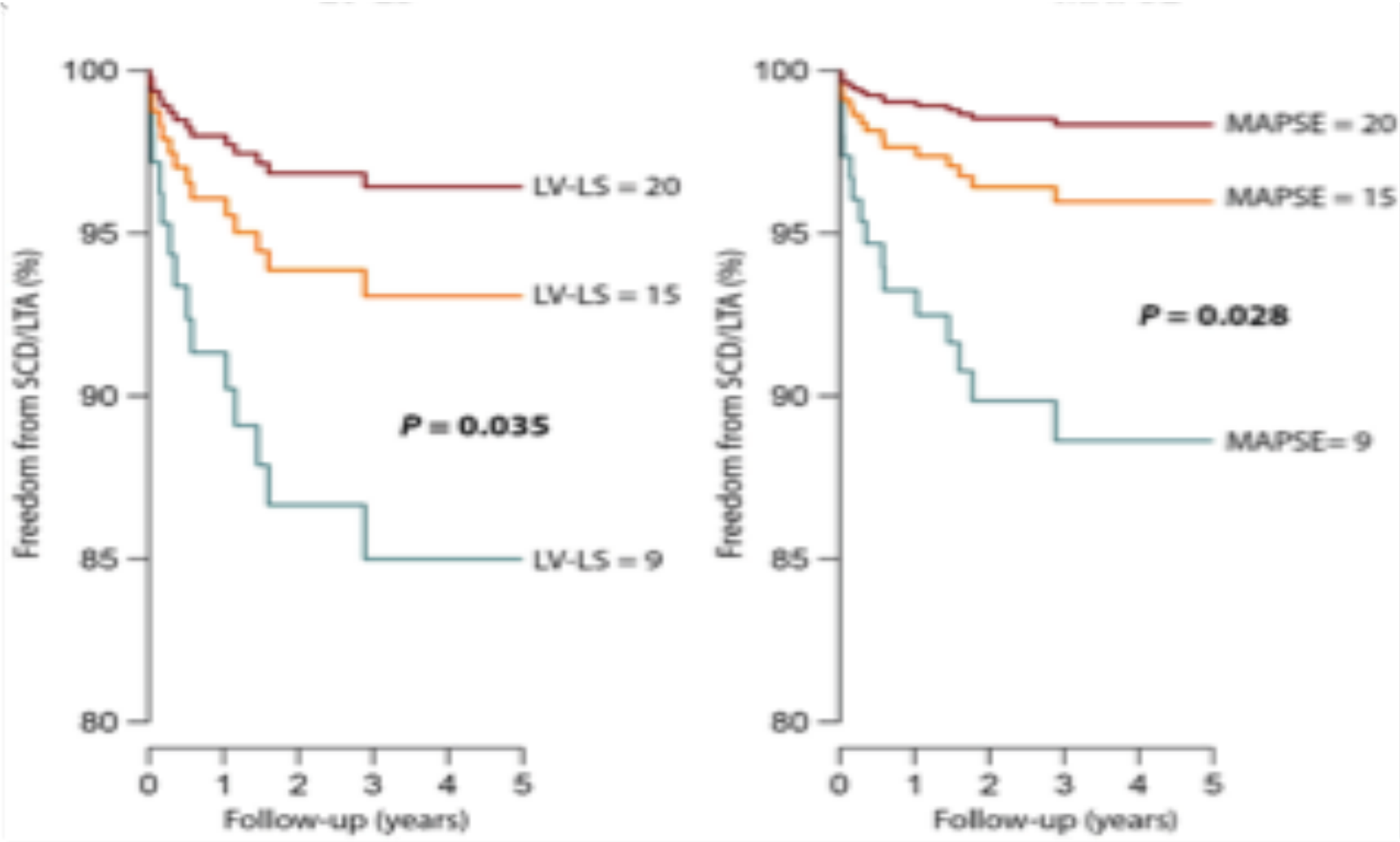


JVP



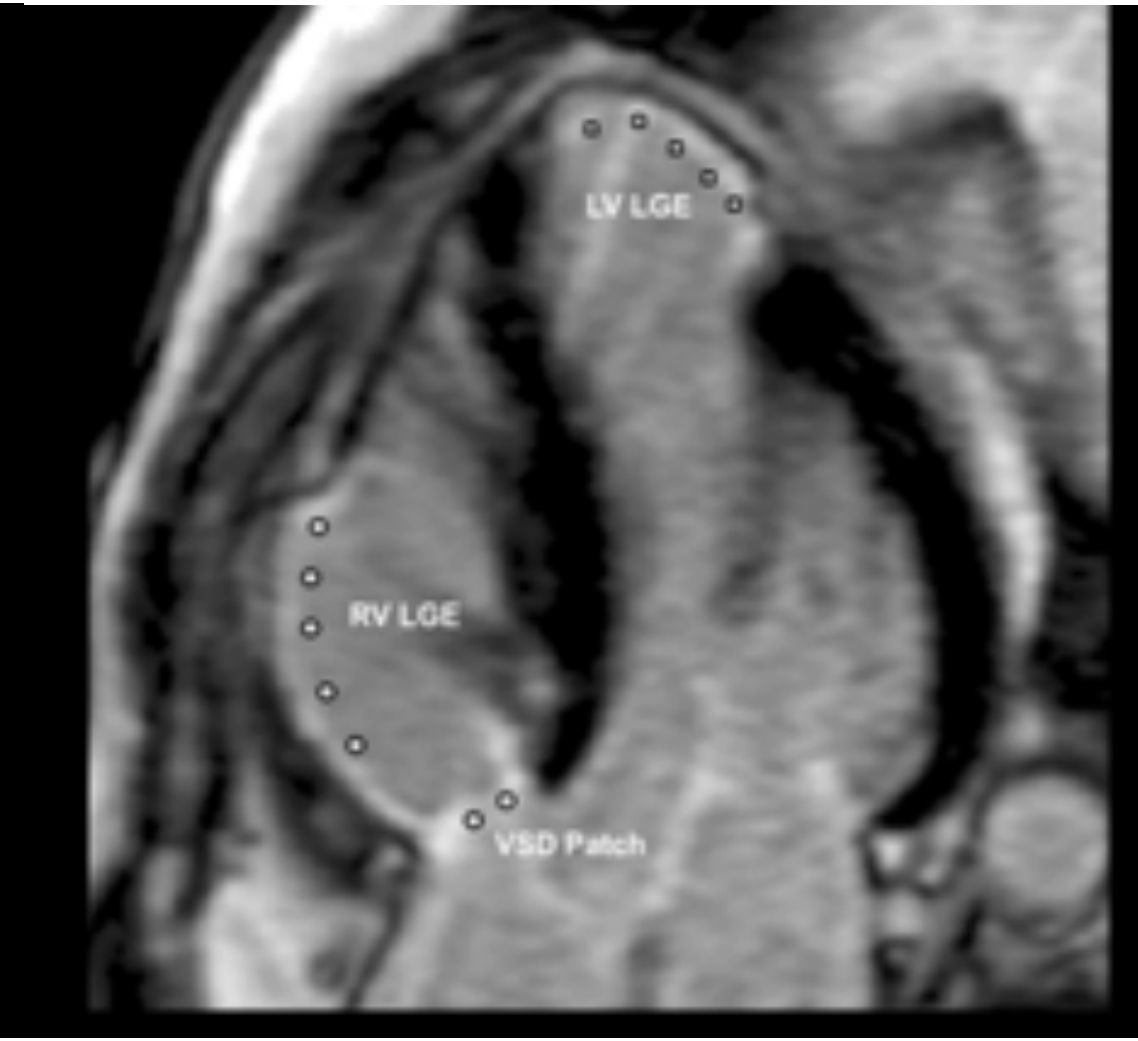
IVC Flow

Survivor LV Function for freedom from SCD + Life Threatening Arrhythmia



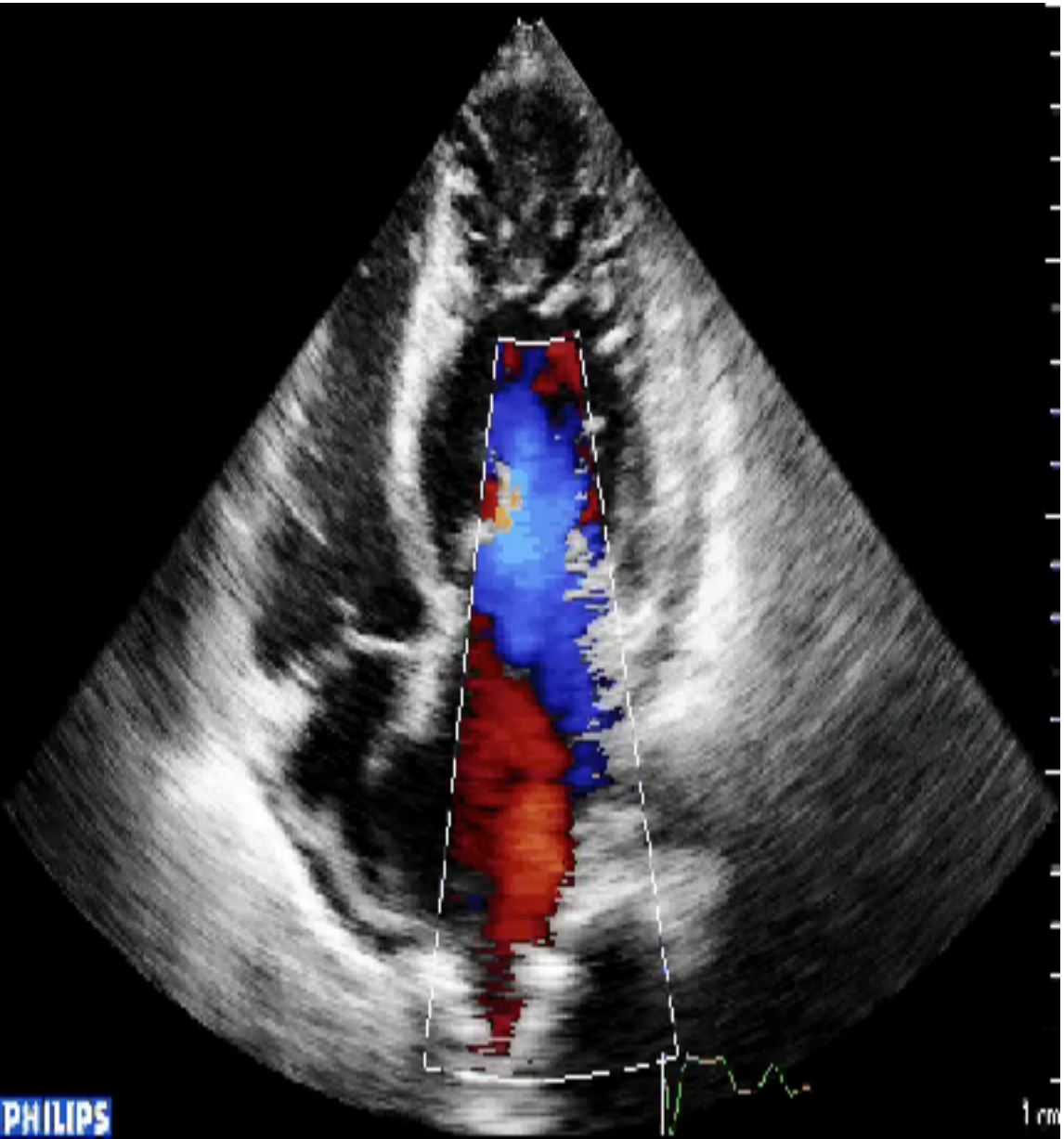
Diller GP et al, Circulation 2012,125:2440-2446

Repaired Fallot - Contrast Echo for detecting fibrosis



S. Bhattacharyya et al. / International Journal of Cardiology 166 (2013) e16–e18

Congenitally Corrected TGA



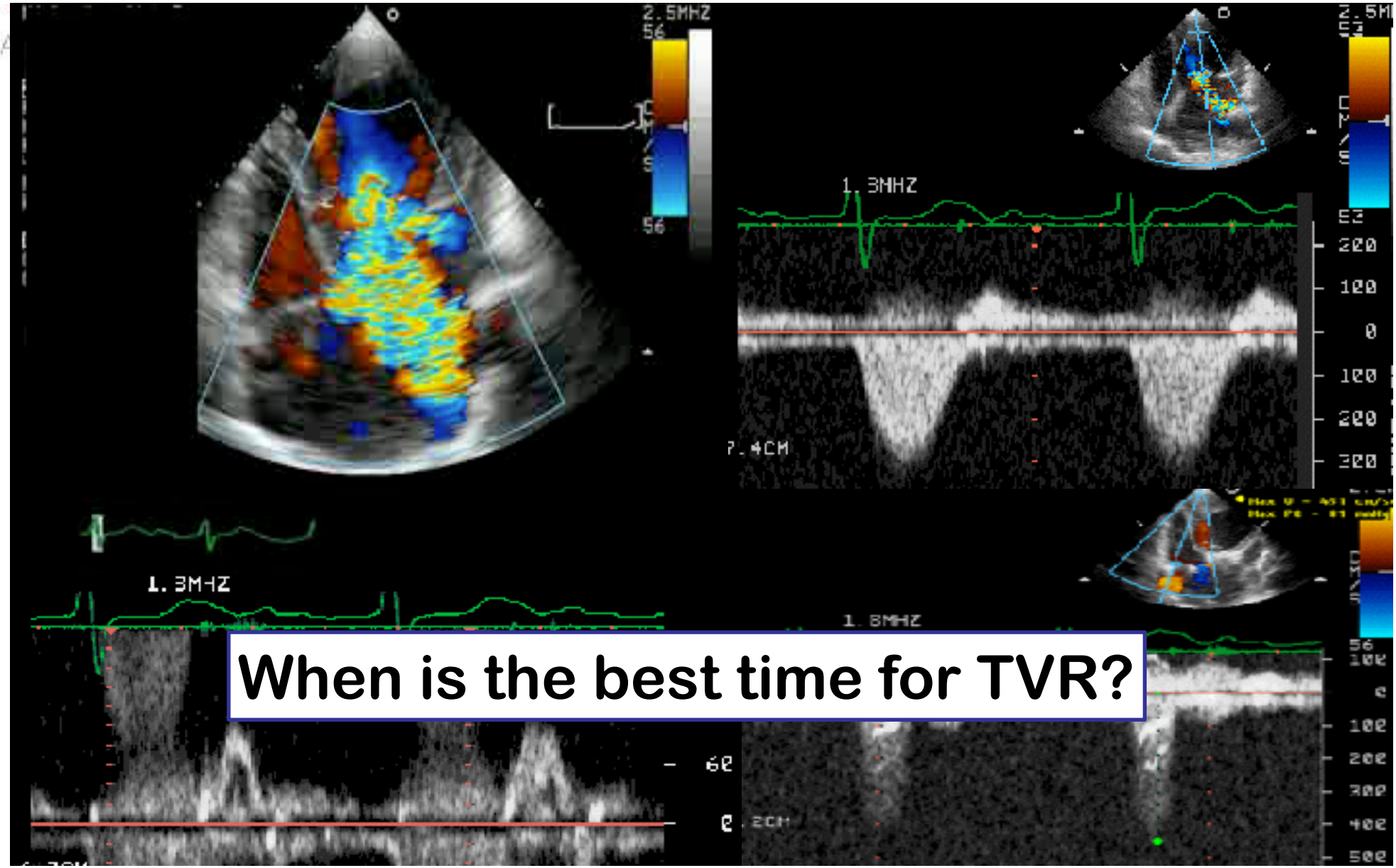
Associated lesions:

- VSD, PS
- Ebstein anomaly of TV
- Heart block

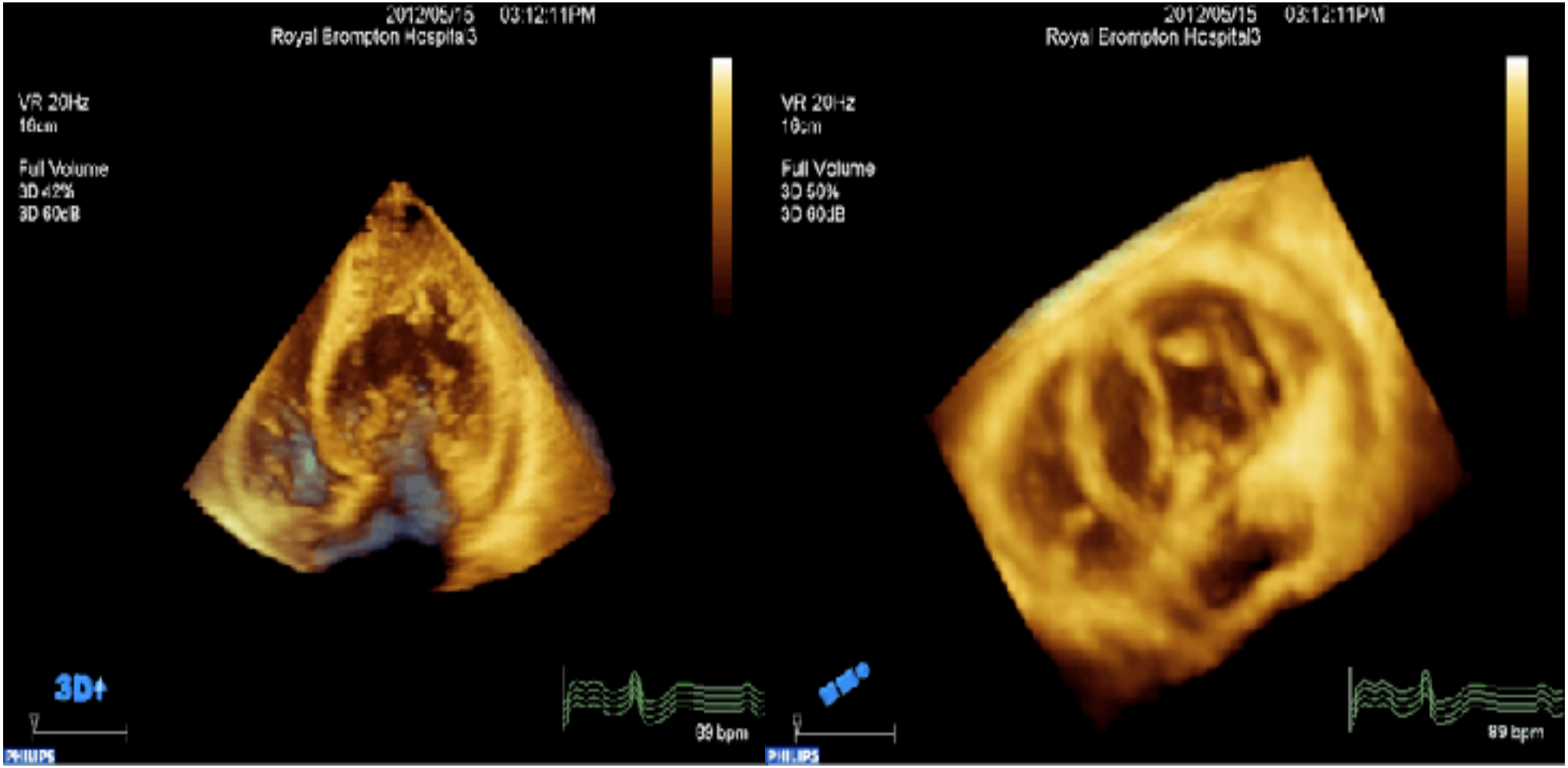
Issues:

- Systemic RV dysfunction
- Tricuspid valve regurgitation
- Conduit stenosis (after LV-PA conduit repair)

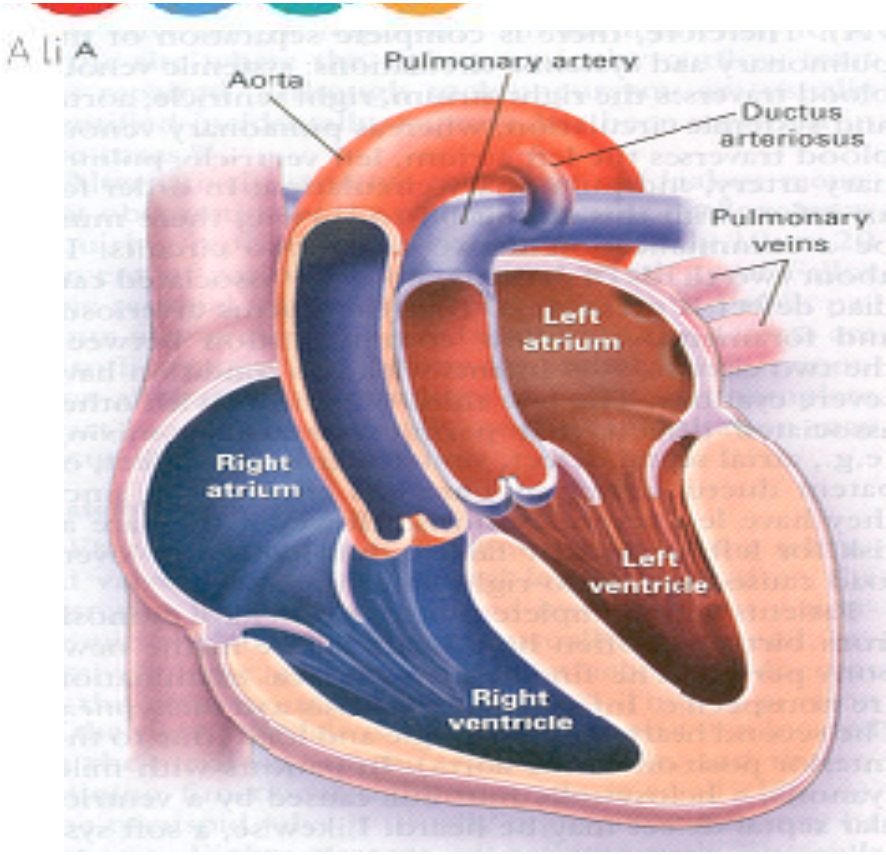
CC - TGA with severe TR

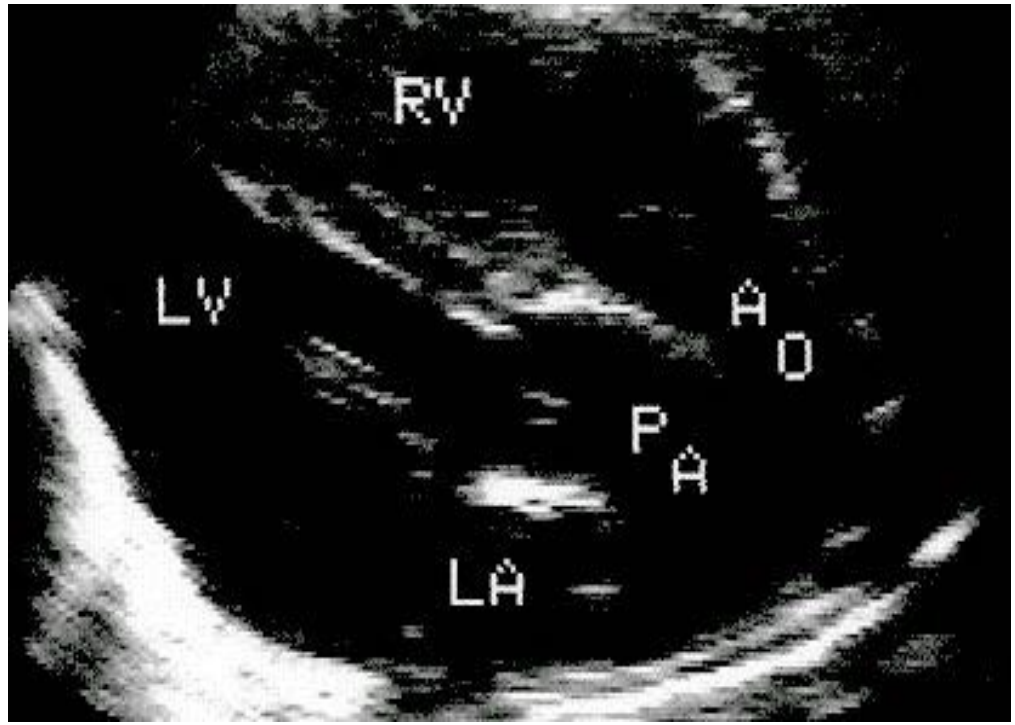


CC-TGA with Quadra-cuspid left AV valve



Transposition of Great Arteries

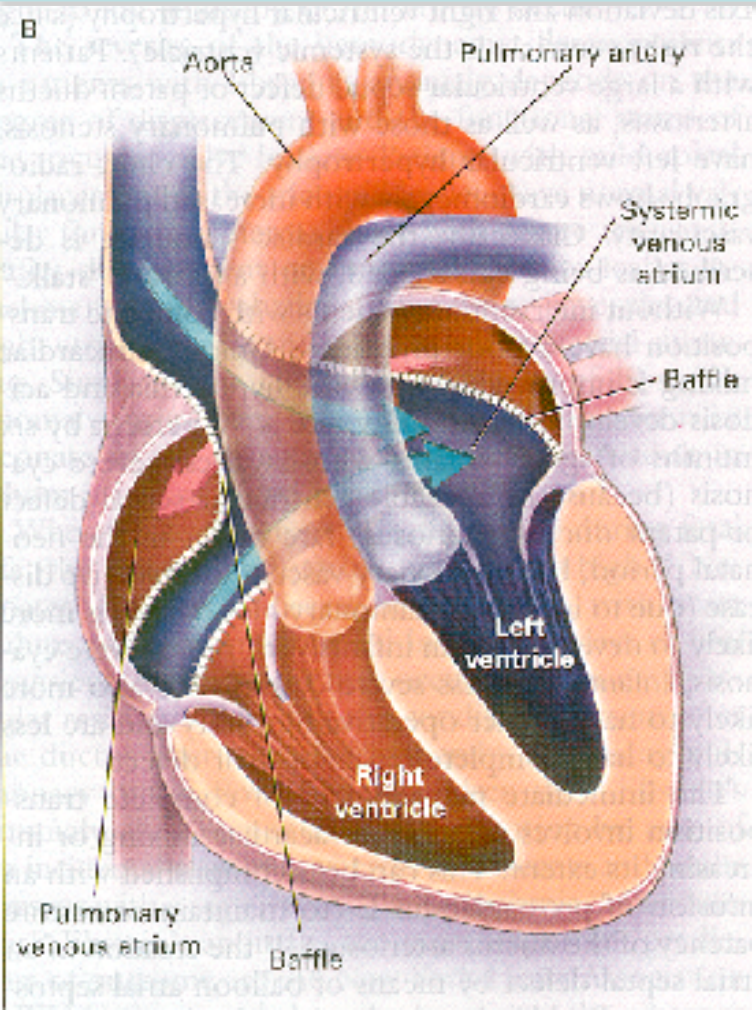




Associated abnormalities:
VSD
LVOTO (PS)
Variable coronary artery anatomy

Surgical repair
1. Mustard or Senning operation
2. Arterial Switch operation
3. Rastelli type repair

The Issues - TGA post Atrial Switch Repair

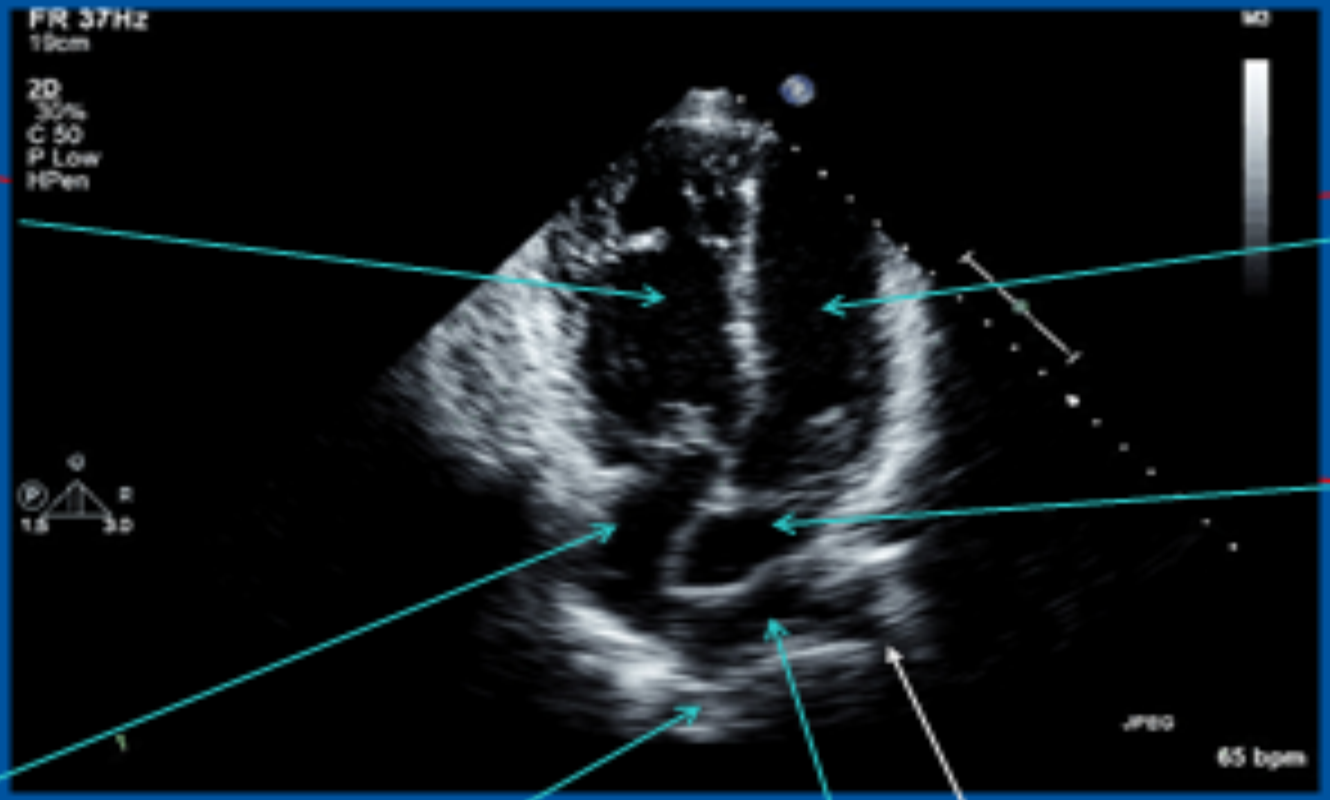


- Systemic RV dysfunction
- Tricuspid valve regurgitation
- Venous baffle obstruction and baffle leak
- Pulmonary hypertension
- Residual VSD
- Arrhythmia:

  bradyarrhythmias and tachyarrhythmias

2D echo assessment: apical 4Ch view

Universitair Medisch Centrum Utrecht



Right ventricle

Left ventricle

Left atrium

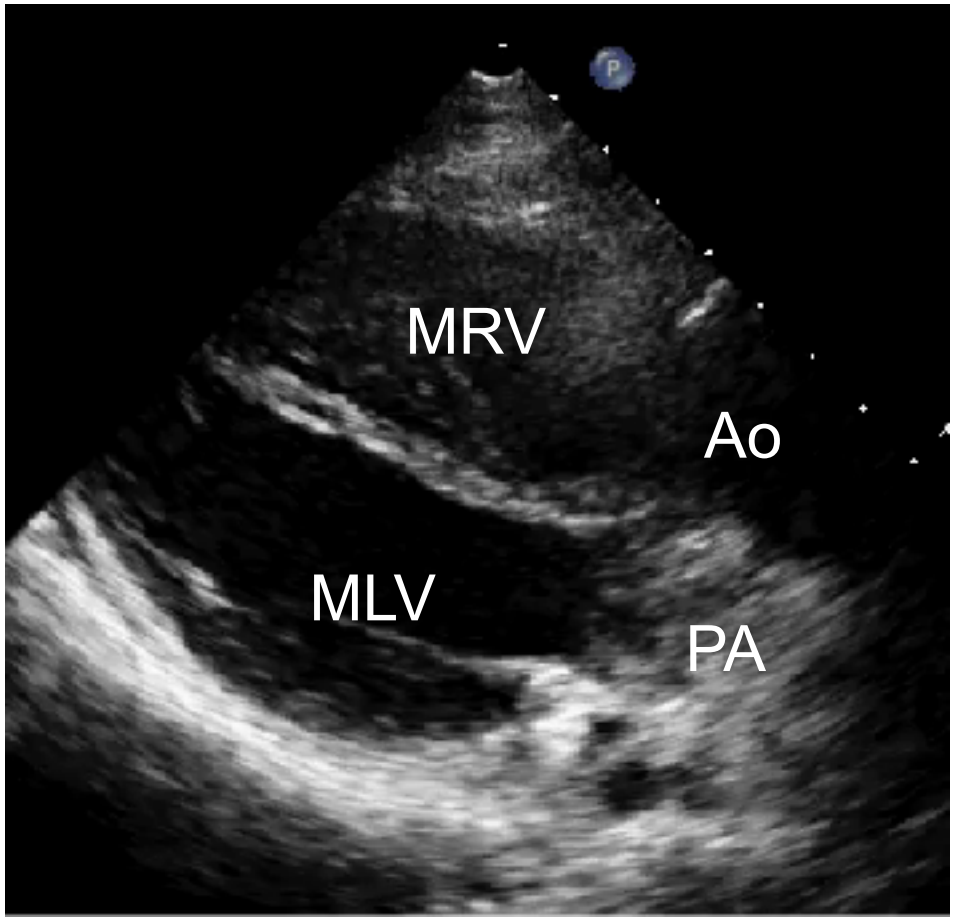
Right atrium

right pulmonary veins

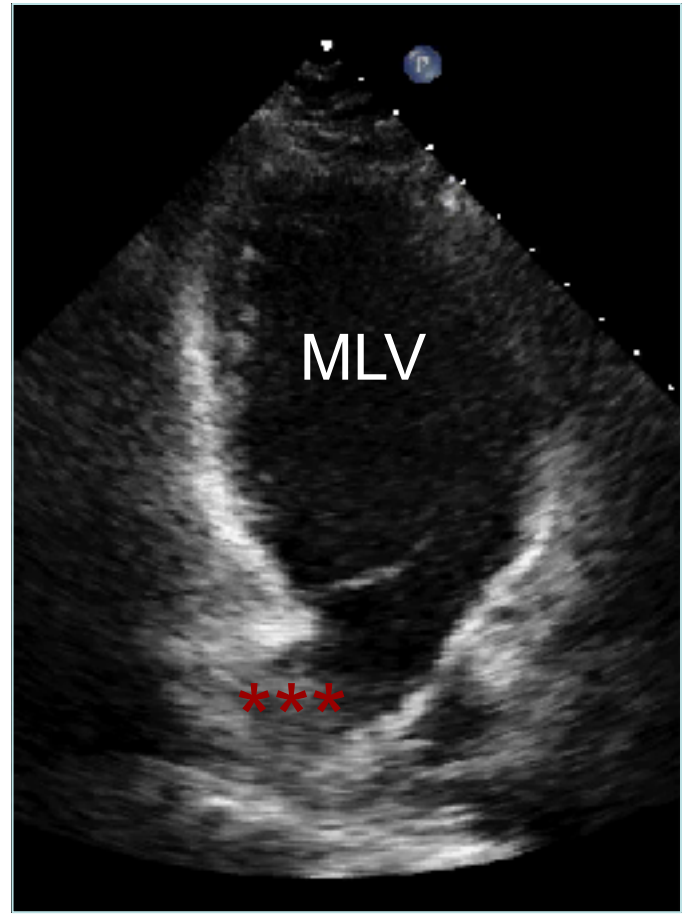
Left pulmonary veins

pulmonary venous atrium

TGA after Mustard / Senning Repair



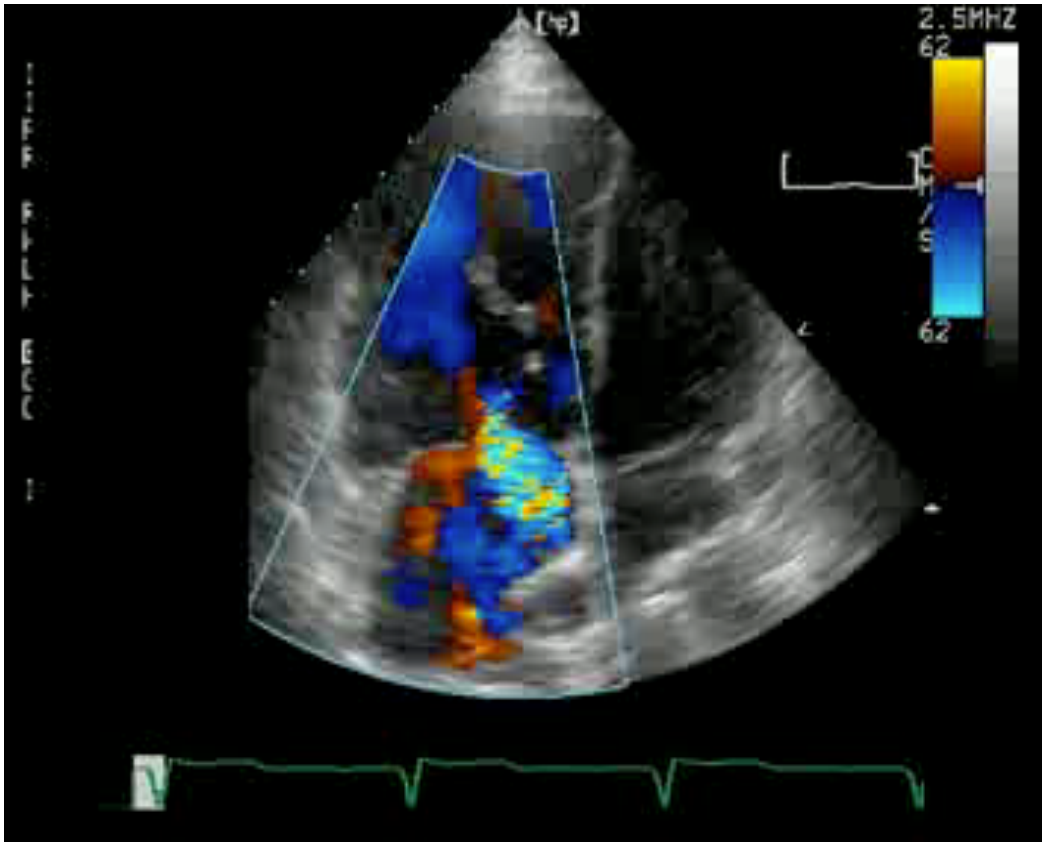
Parasternal long axis view

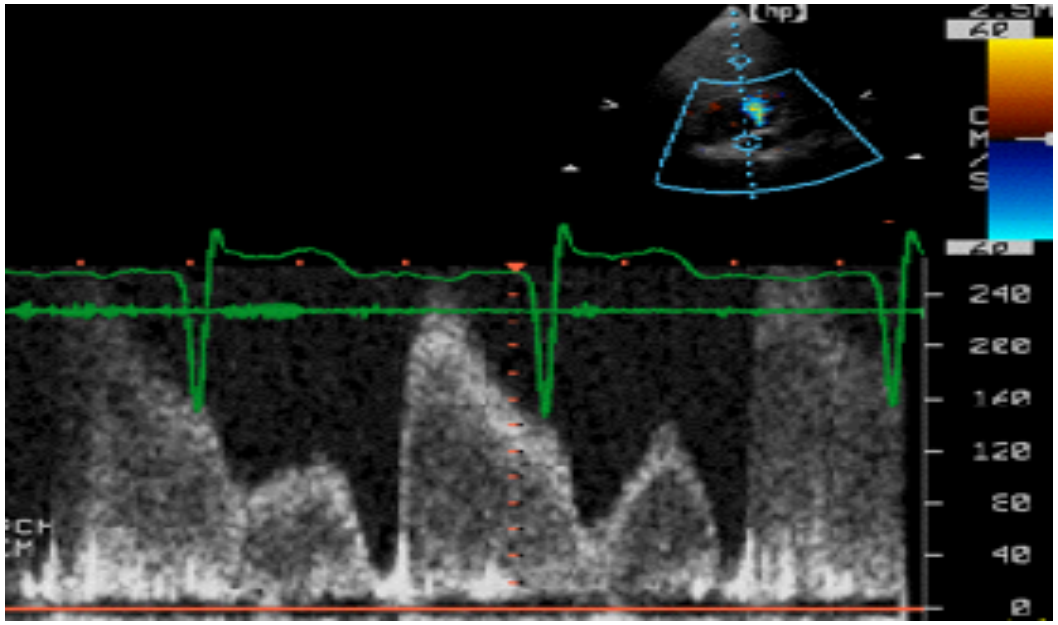


Apical two chamber view

*** IVC pathway

The Adult with Mustard for TGA
Baffle Obstruction

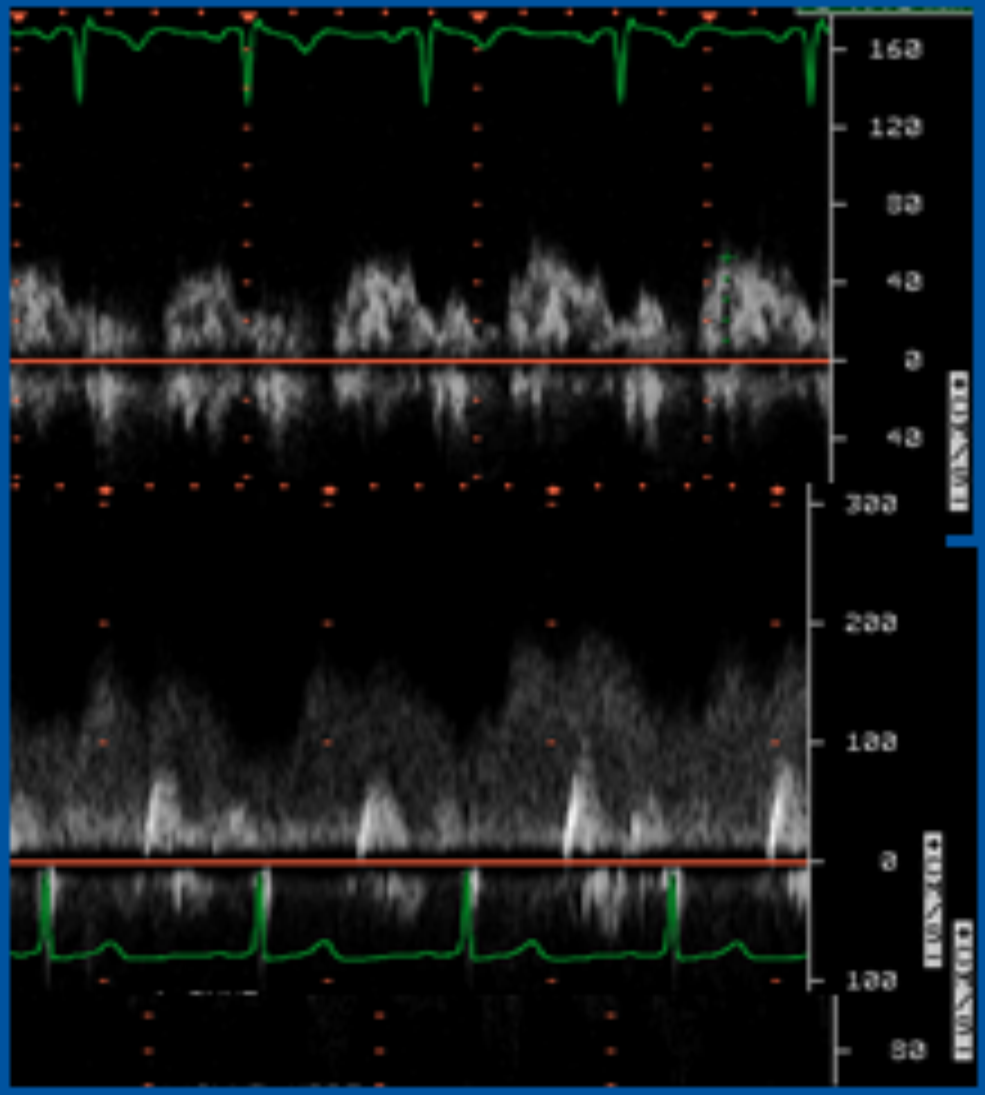




Doppler flow of Pulmonary venous pathway

Pulsed wave Doppler in tunnels

Universitair Medisch Centrum Utrecht



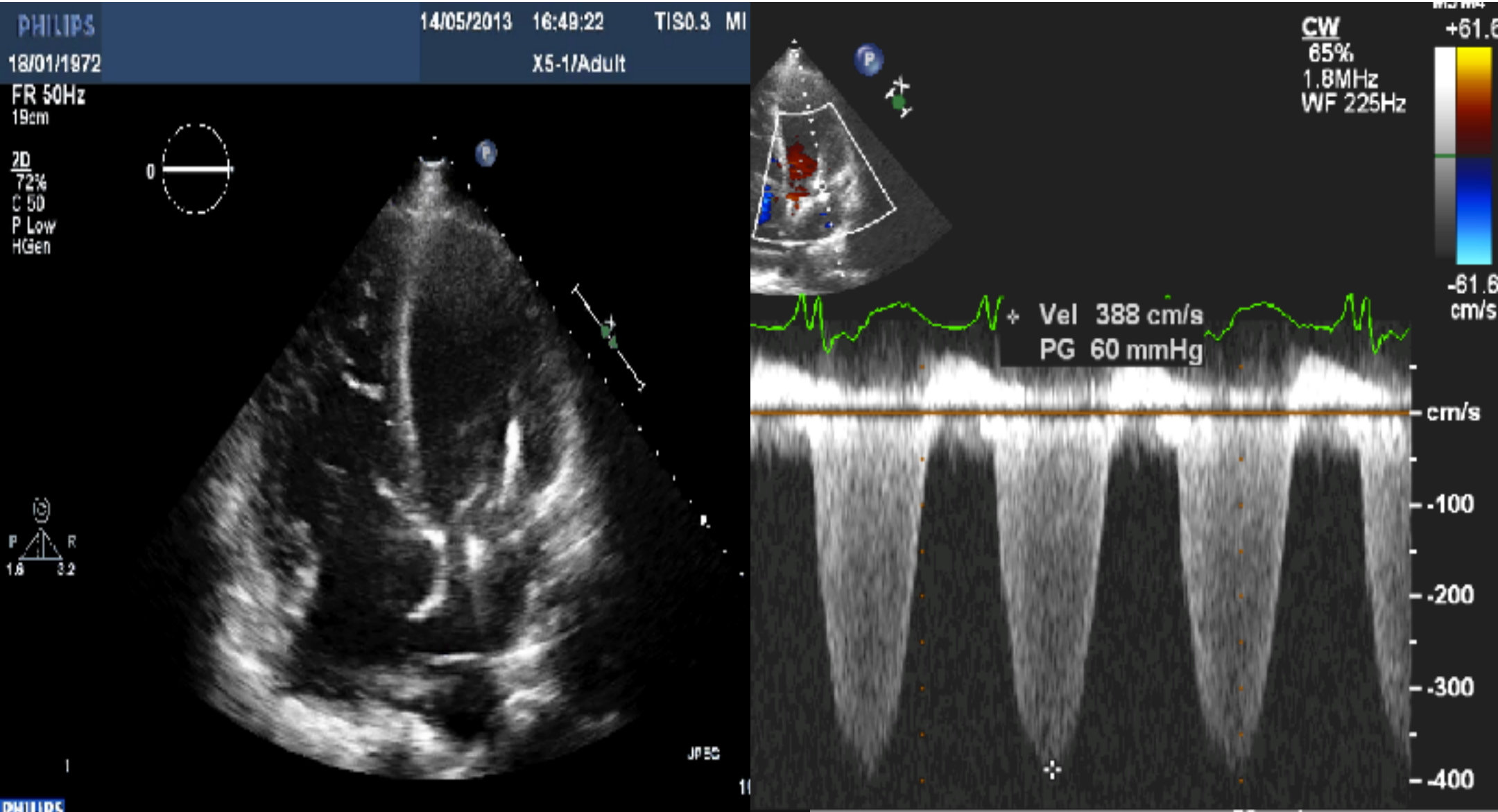
Phasic flow, S<D,
proximal in tunnel

More distal in tunnel:
1.8 m/s, continous flow

→ severe stenosis

**Not (only) velocity;
flow pattern counts most**

The Adult with Mustard for TGA
PAH and BiV Failure

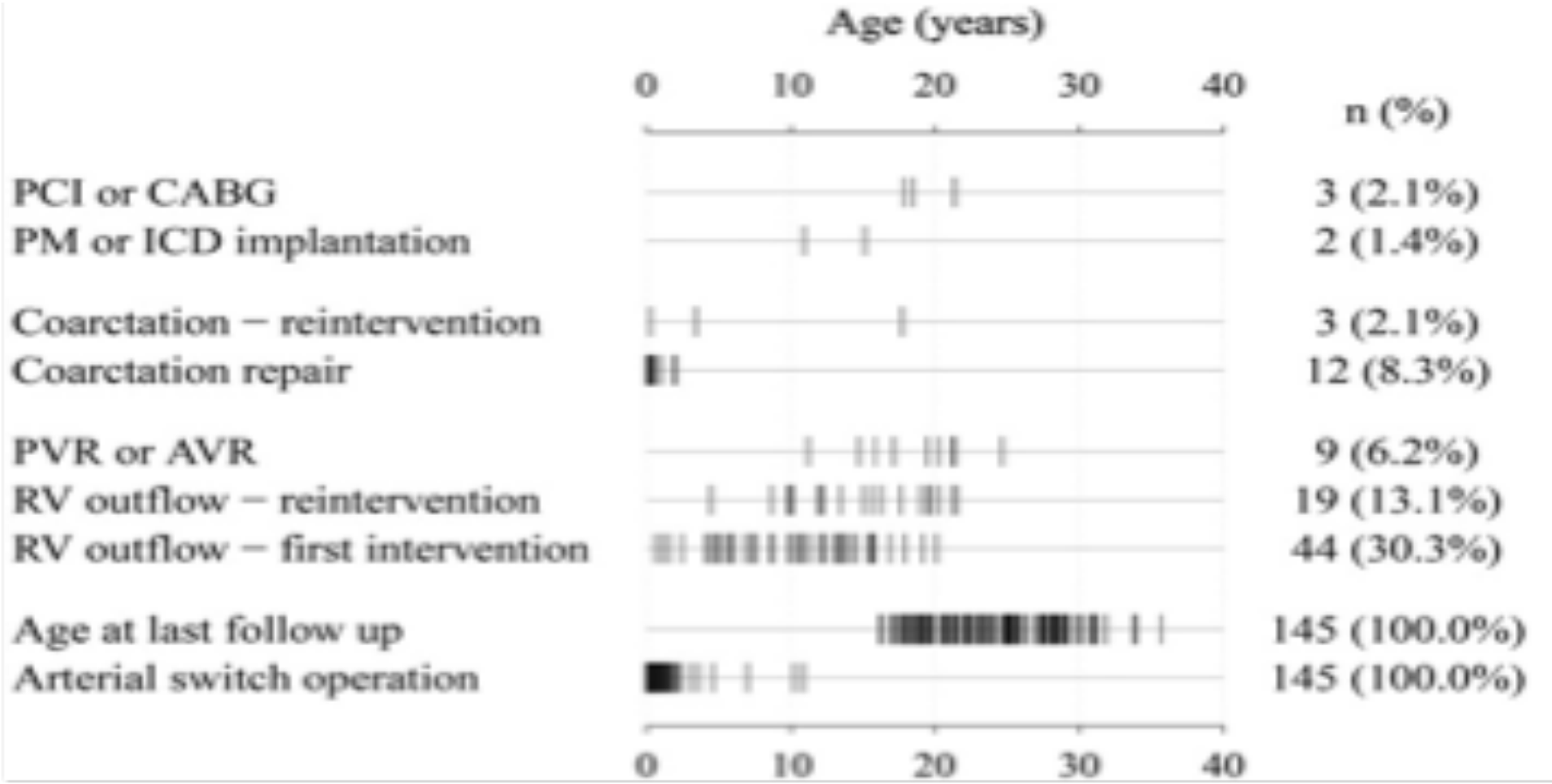


The Issues - TGA post Arterial Switch Repair

- Pulmonary stenosis: mainly supravalvar or peripheral PS
- Neo-aortic valve regurgitation and aortic root dilatation
- Coronary artery anomaly: stenosis or occlusion
- Biventricular dysfunction

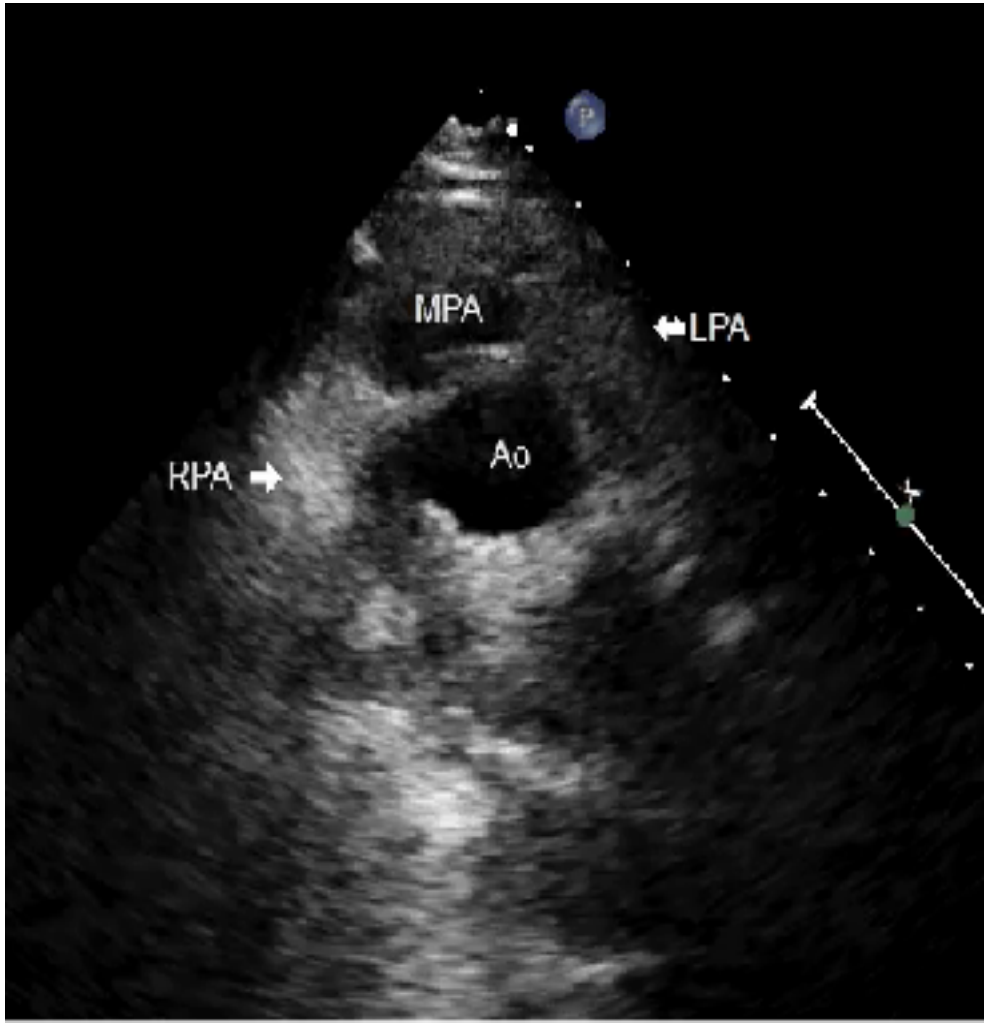
But, systemic LV is preserved

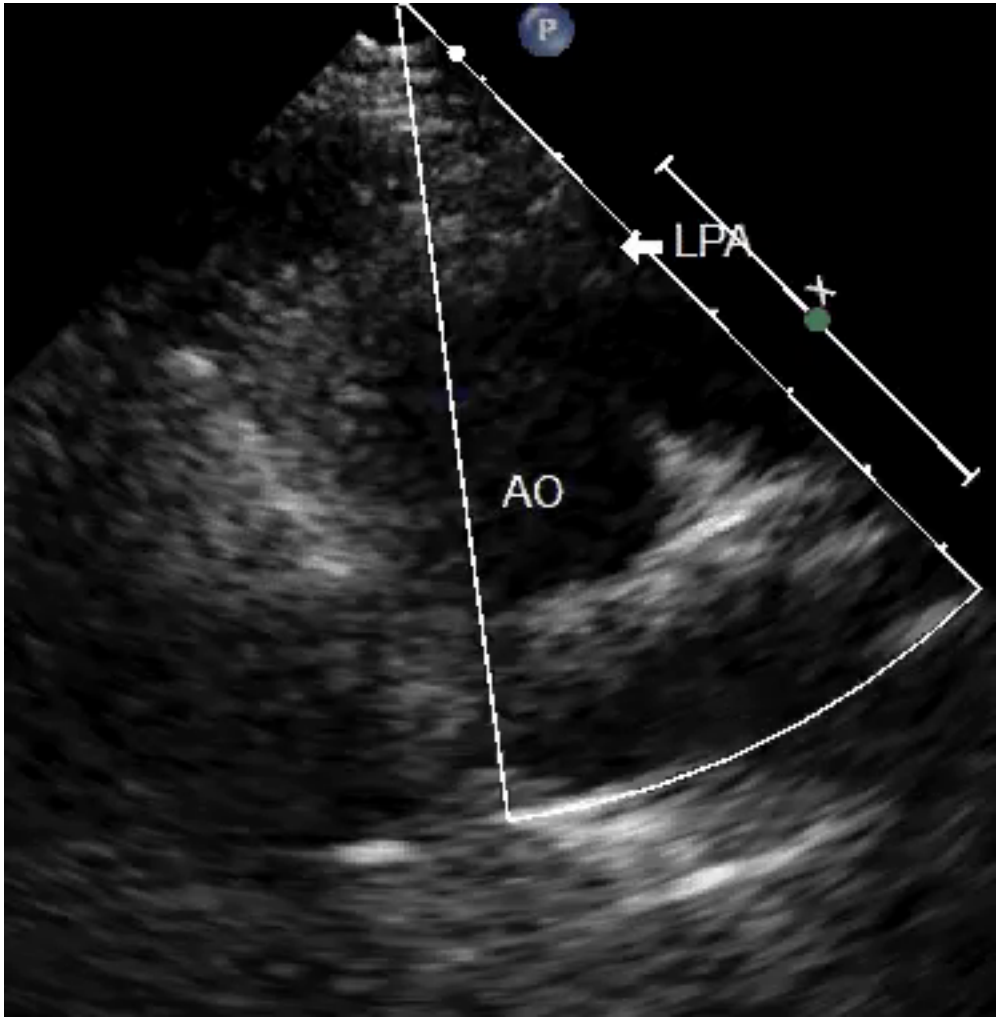
Type of Interventions and distribution In pts after Arterial Switch operation



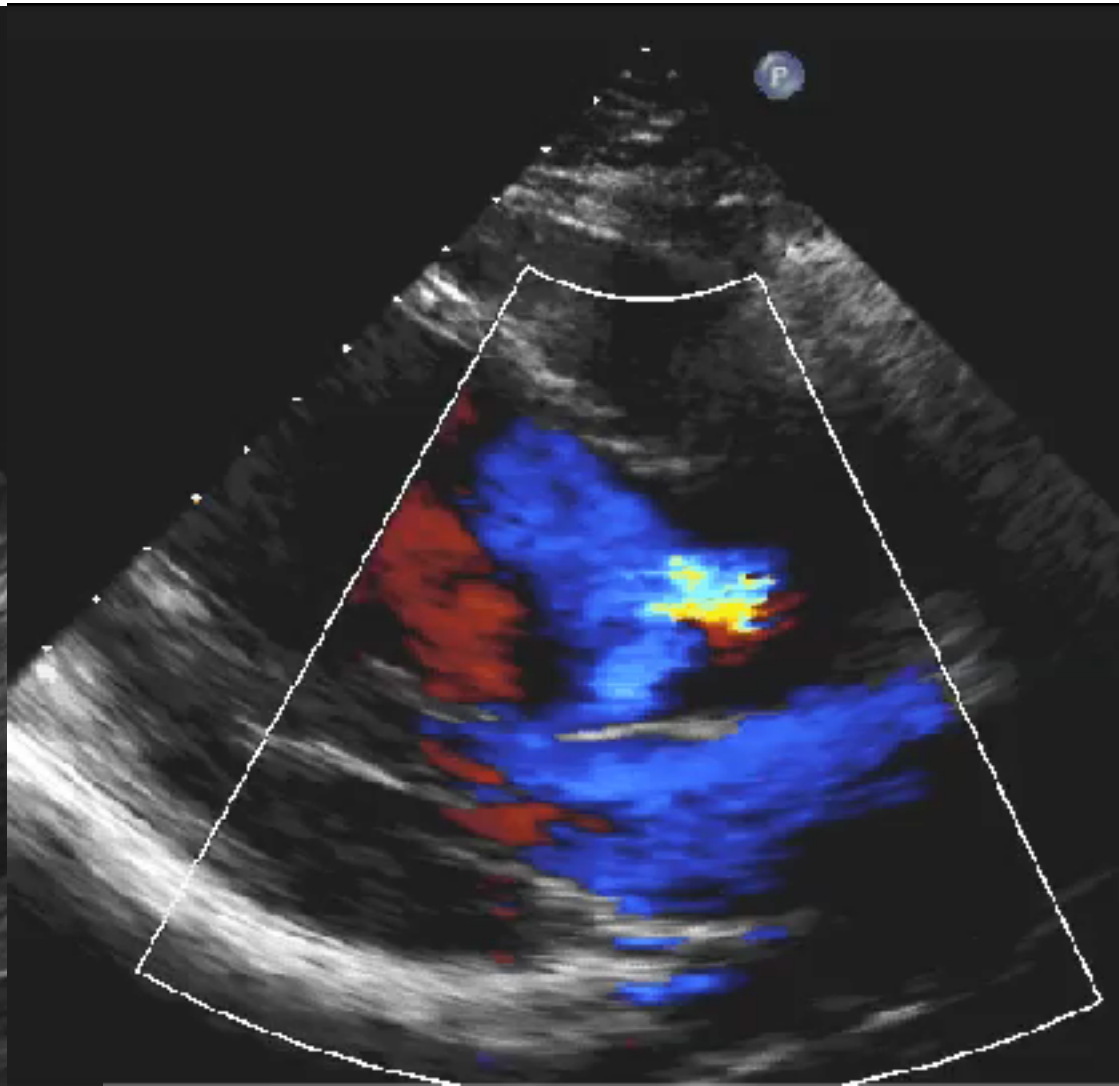
Kempny A, et al. IJC (2012), doi 10, 1016

Branch PA stenosis after Arterial Switch Repair





TGA after Arterial Switch Operation (ASO)
Late complications - AR



Assessing Univentricular Hearts

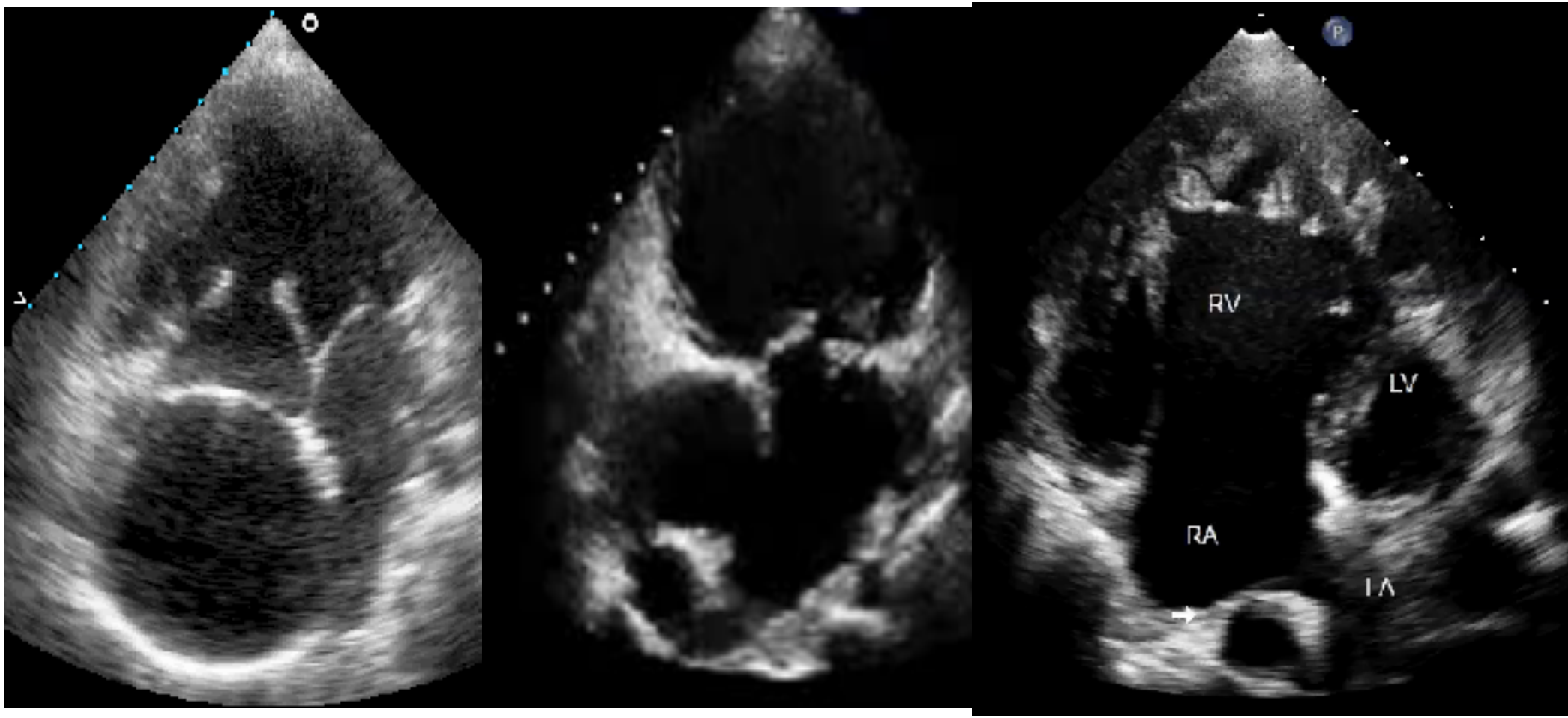
- Ventricular morphology and function
- Atrioventriular valve function
- ? Restriction at atrial level & Size and location of VSD
- Pulmonary stenosis or pulmonary hypertension
- Fontan circuit to exclude:

  obstruction, thrombus, and residual shunts

  sub-AS (restrictive VSD)

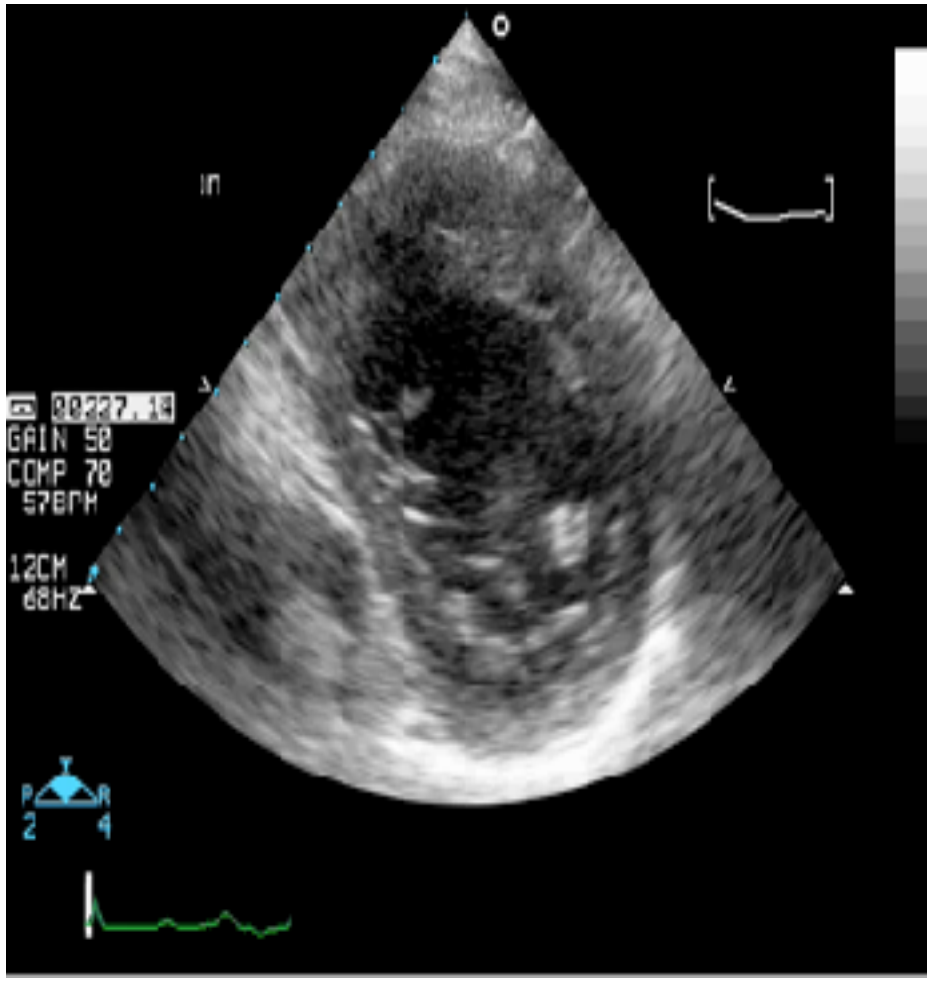
  pulmonary vein compression and obstruction

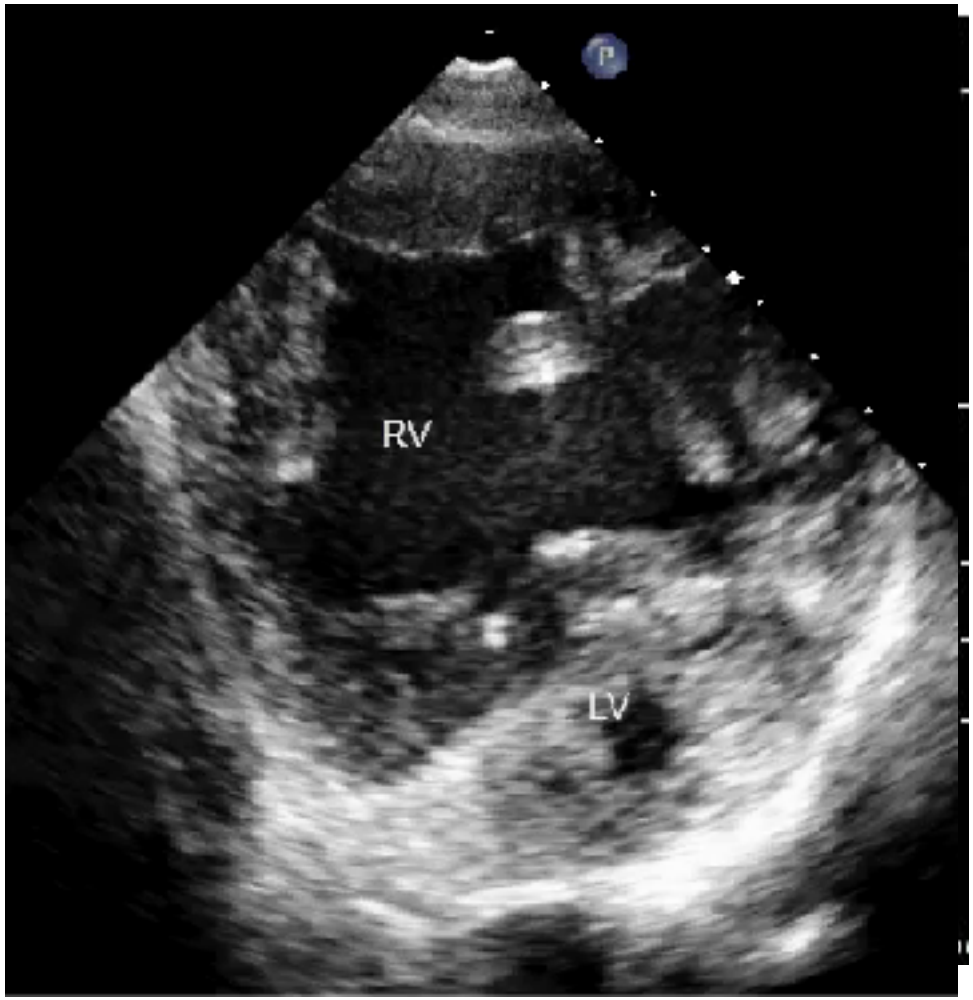
Ventricular Morphology

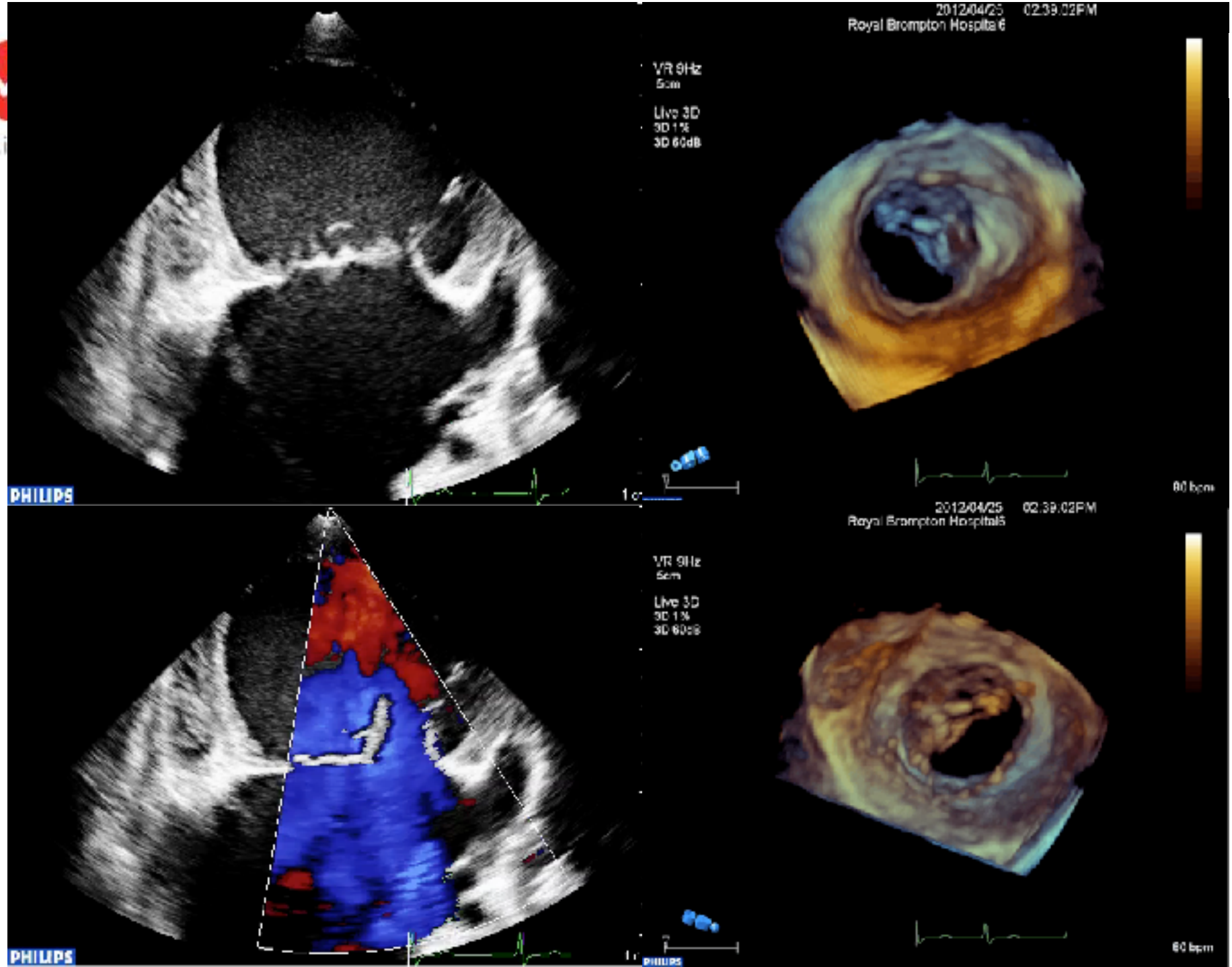


Univentricular atrioventricular connection

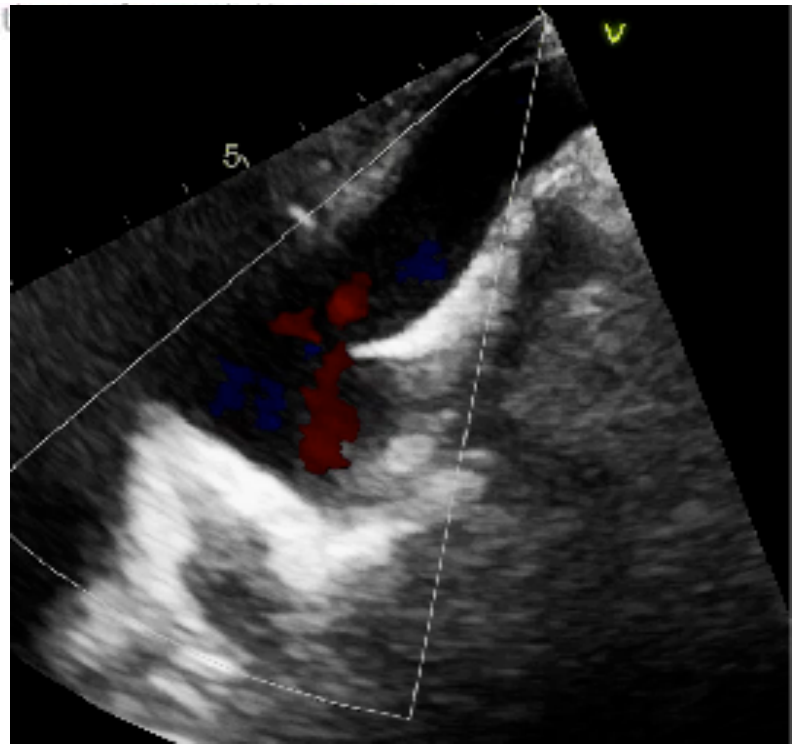
Identification of Ventricles

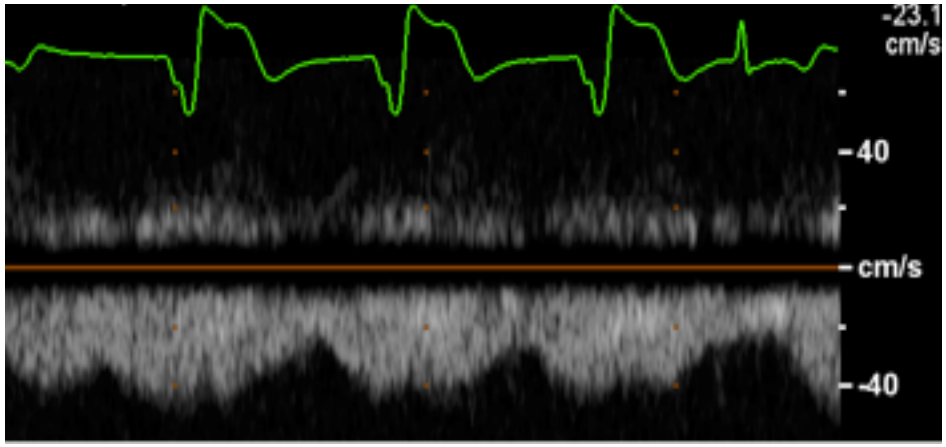





PHILIPS
VR 9Hz
5cm
Live 3D
3D 1%
3D 60dB
2012/04/25 02:39:02PM
Royal Brompton Hospital6
80 bpm
PHILIPS
VR 9Hz
5cm
Live 3D
3D 1%
2012/04/25 02:39:02PM
Royal Brompton Hospital5
80 bpm

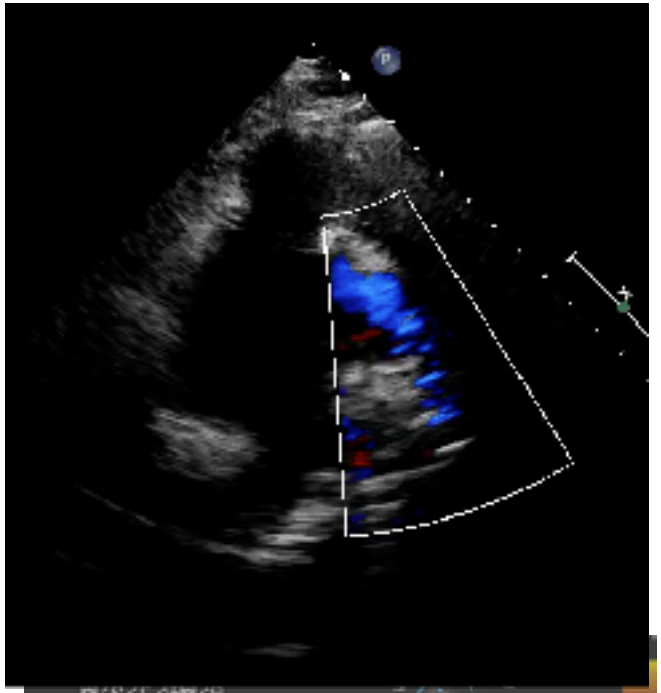
SVC –PA

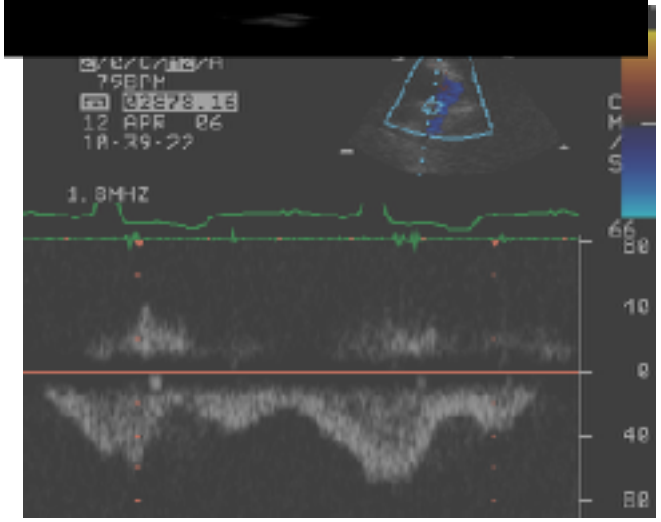


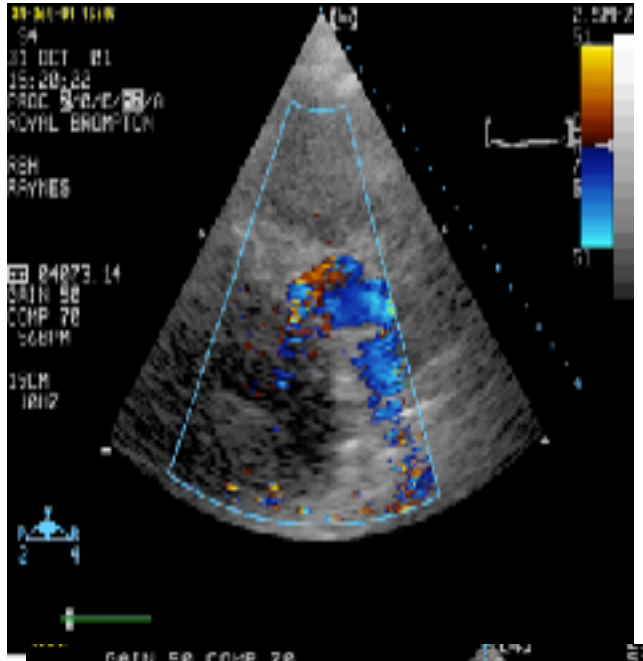


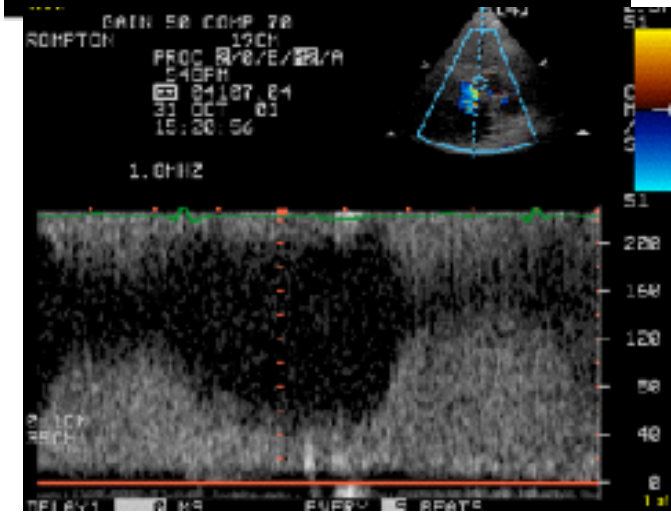
IVC-RA

Fontan Connection

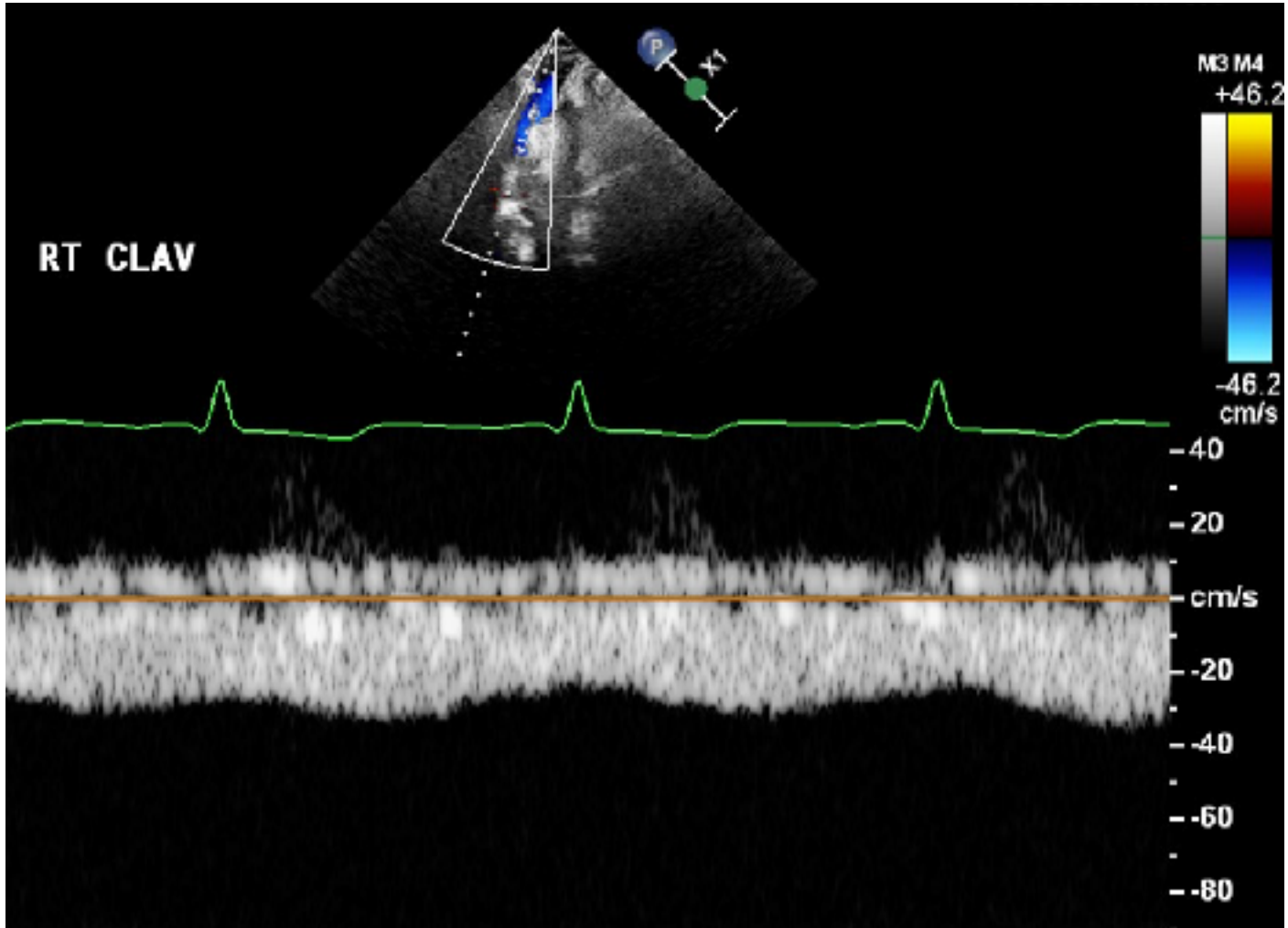




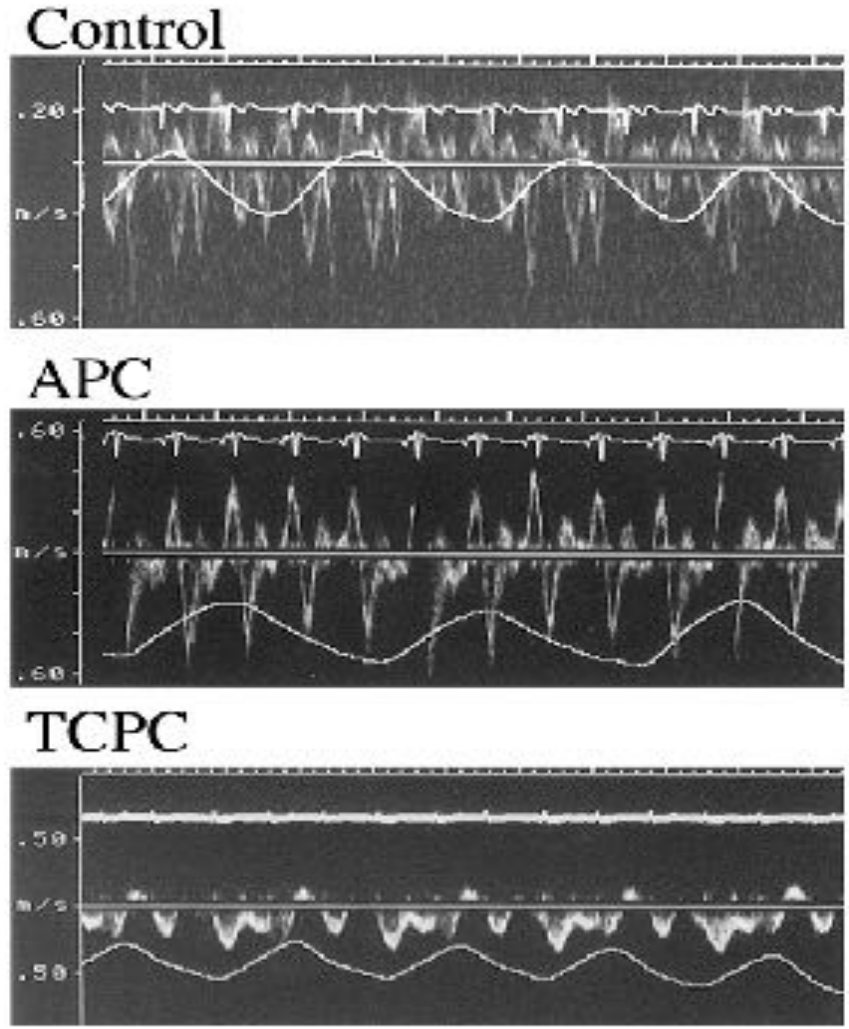




Fontan Obstruction



Pulsed-wave Doppler recordings with simultaneous respiratory monitoring from HV in a control subject, a patient with APC, and a patient with TCPC

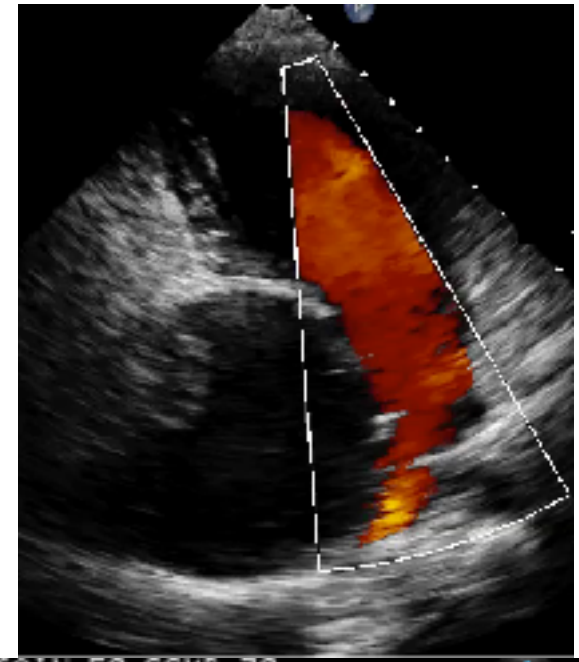


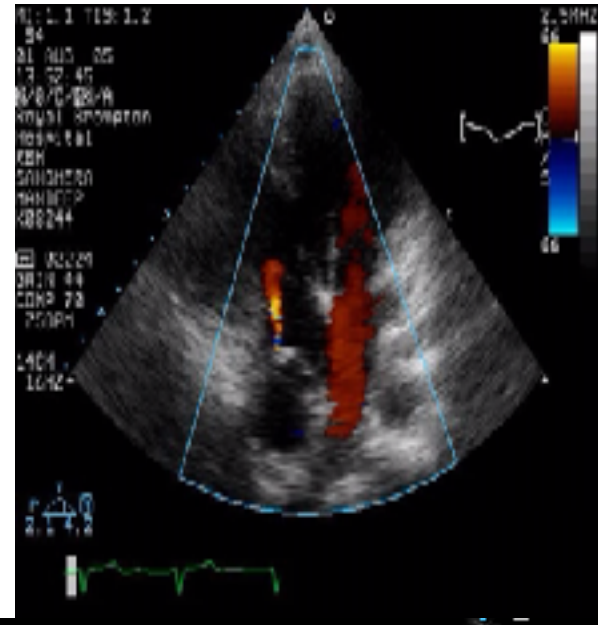
Tain-Yen Hsia et al. Circulation. 2000;102:Iii-148-Iii-153

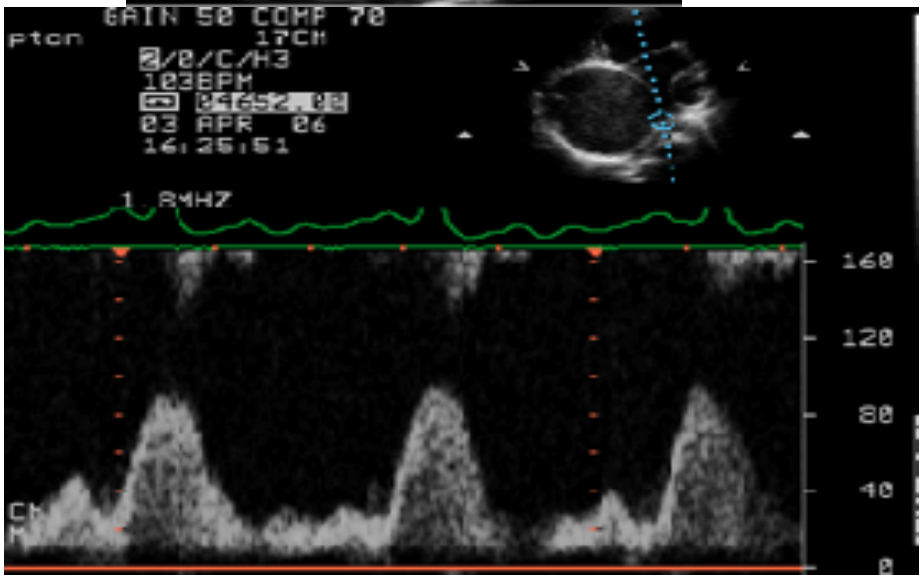
American Heart Association

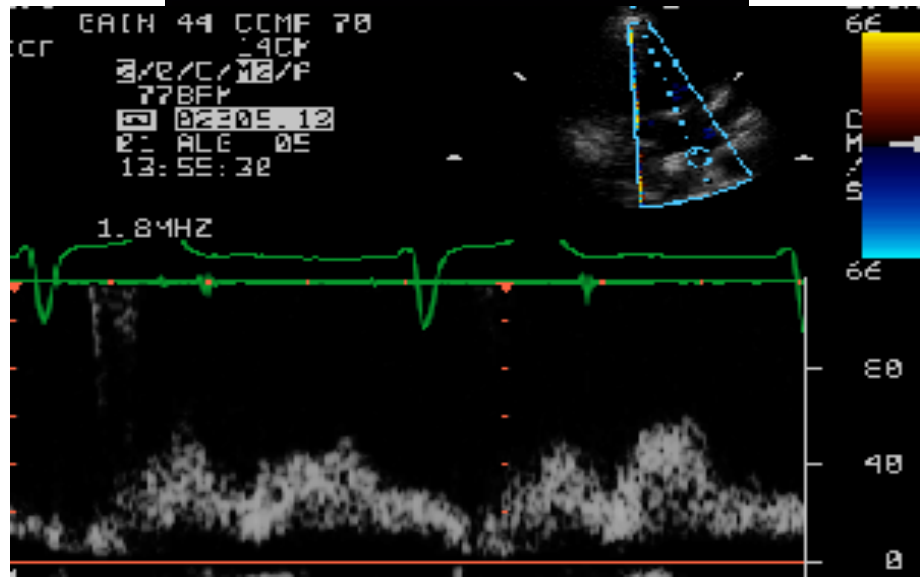
Copyright © American Heart Association, Inc. All rights reserved.

RA dilatation – pulm. venous pathway compression

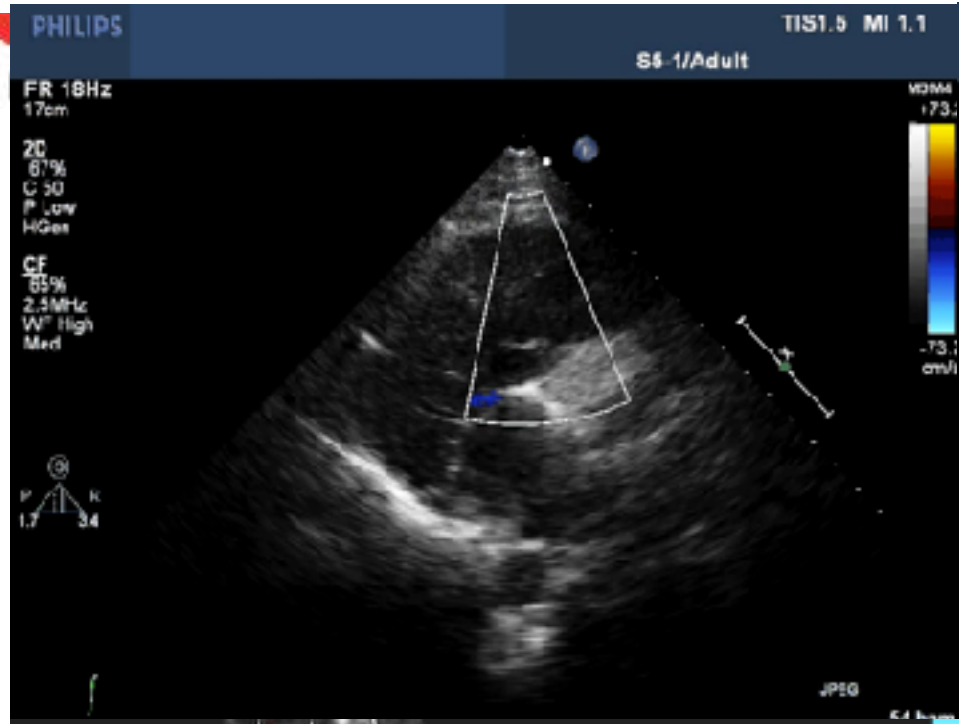


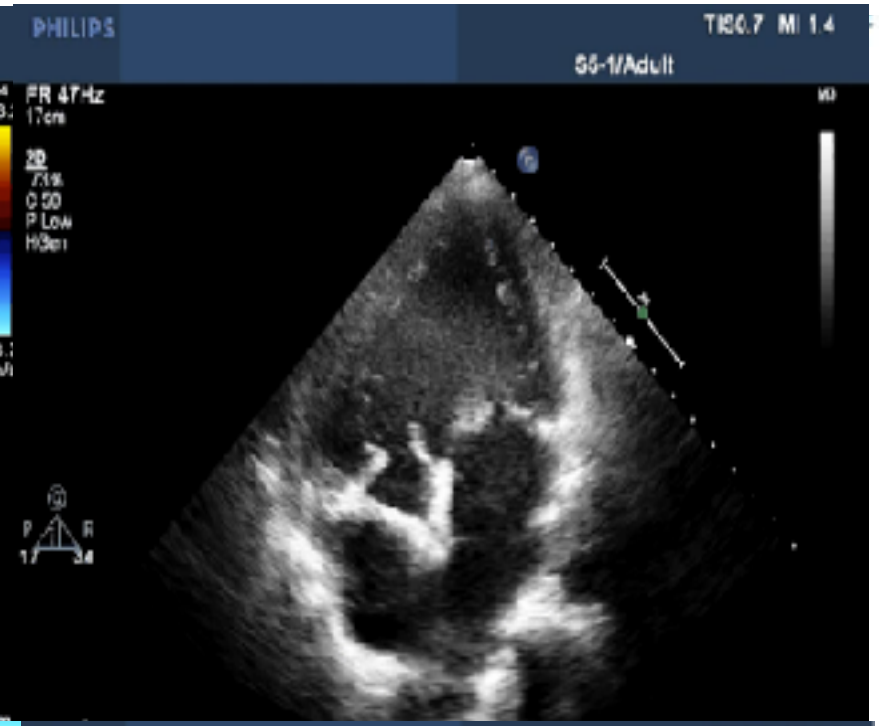


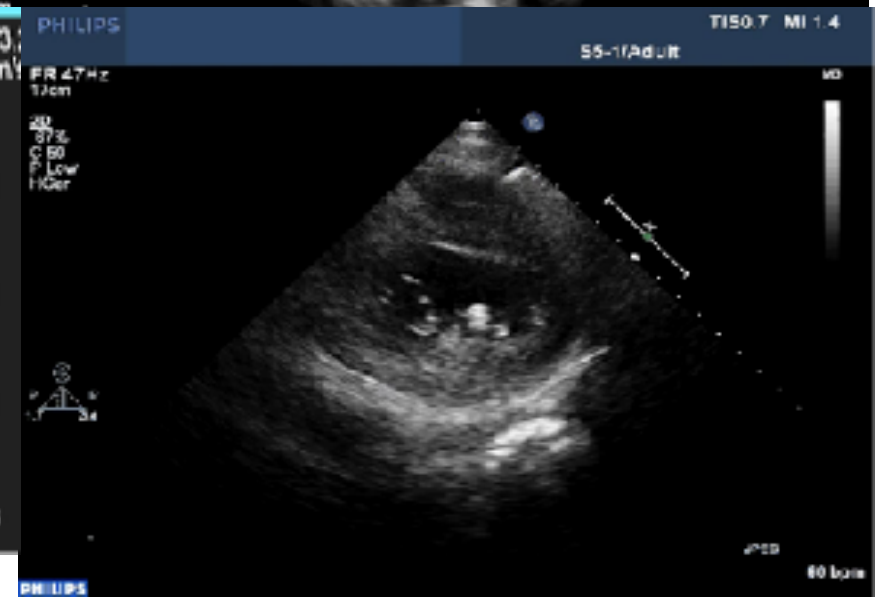




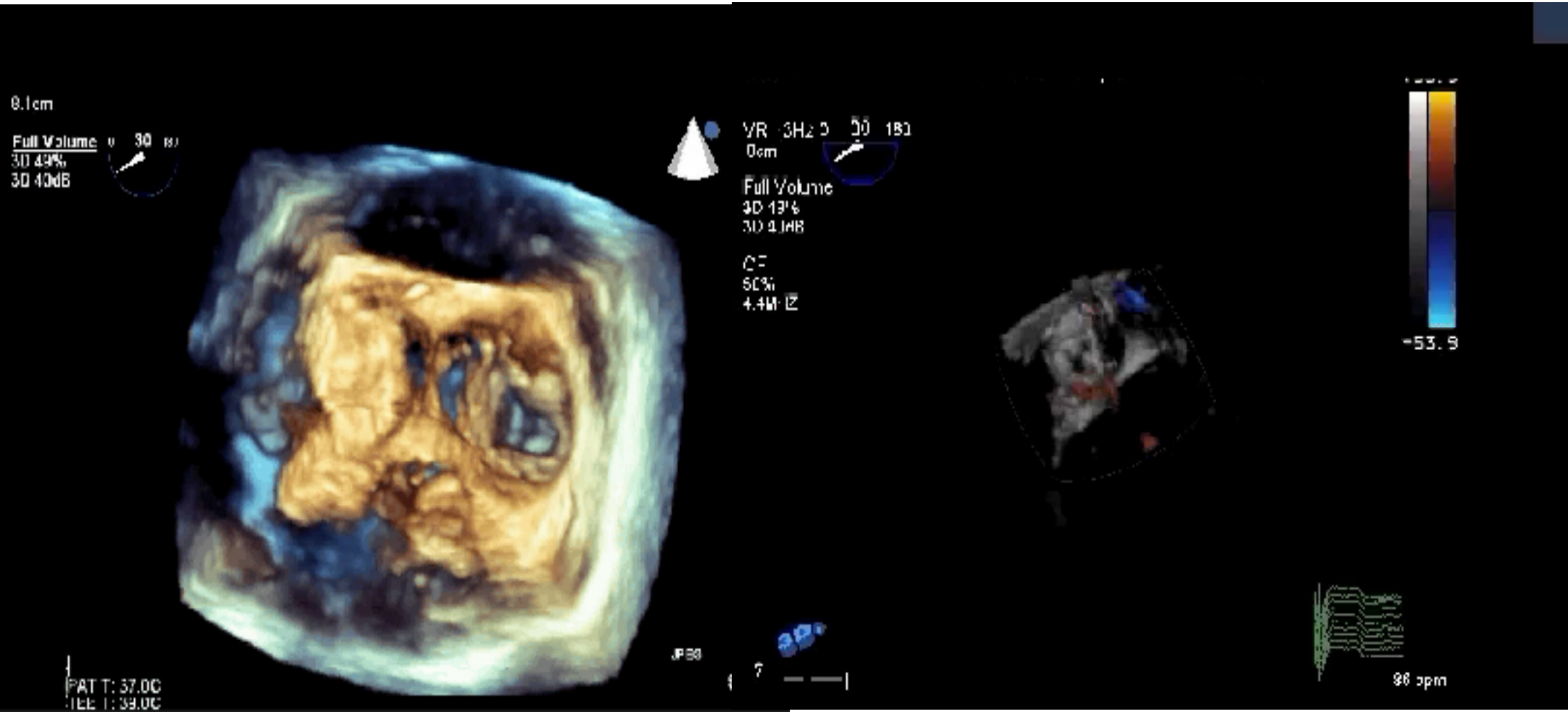
Post Fontan Repair– Restrictive VSD



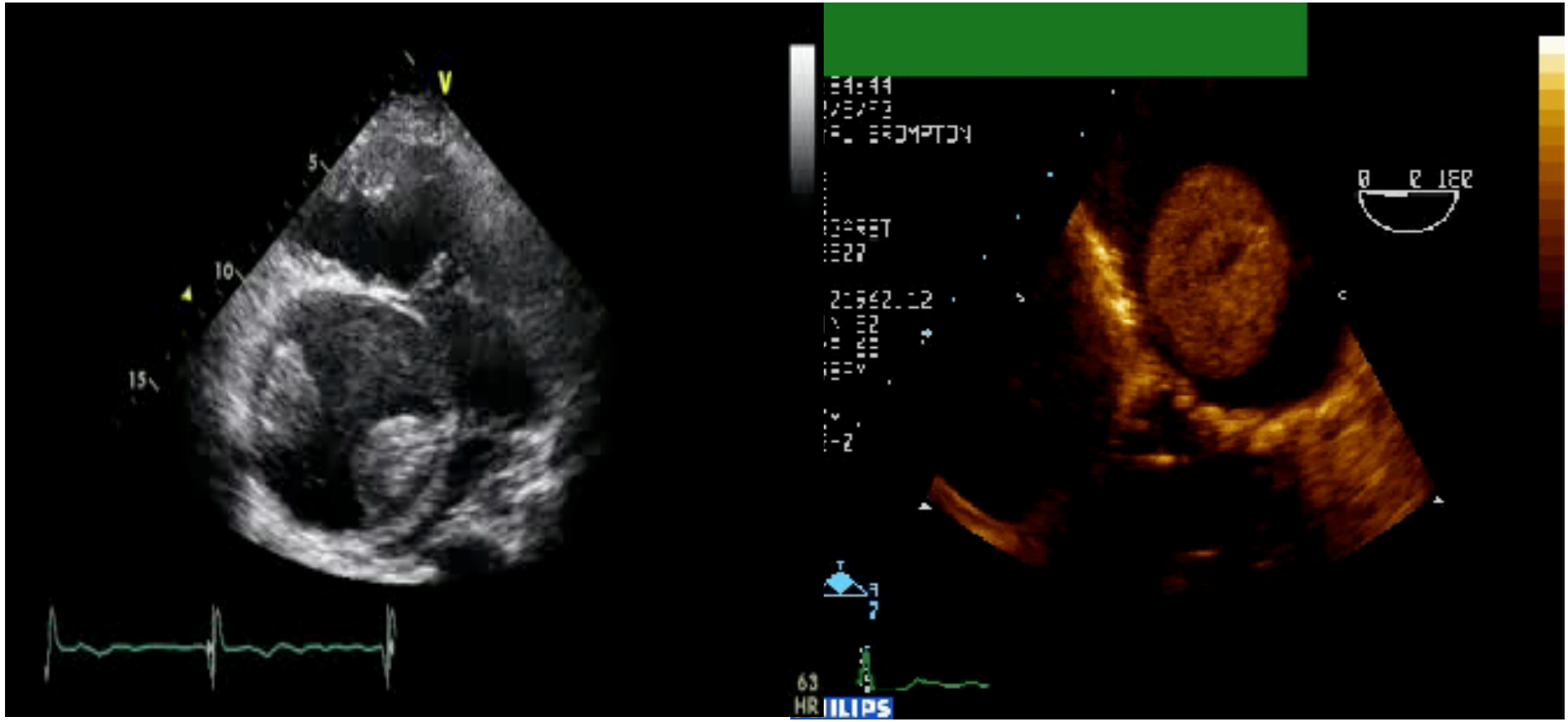




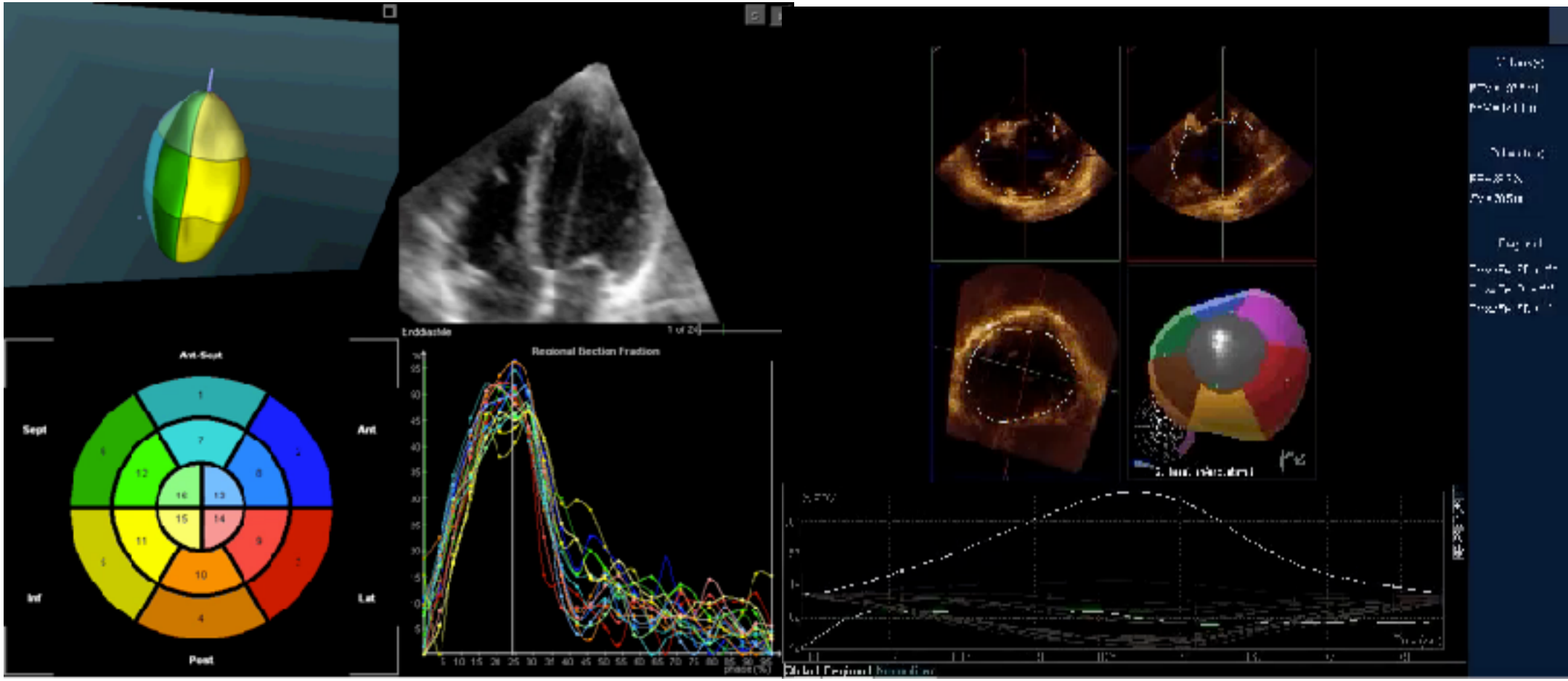
DILV Post Fontan Repair: Patch Leak



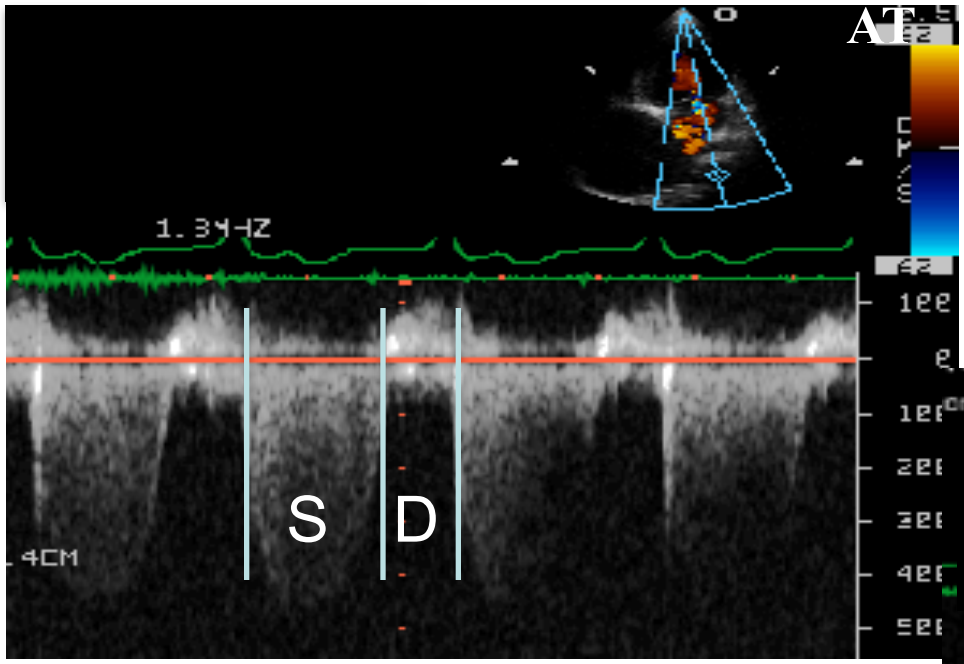
Single Ventricle – Thrombus formation



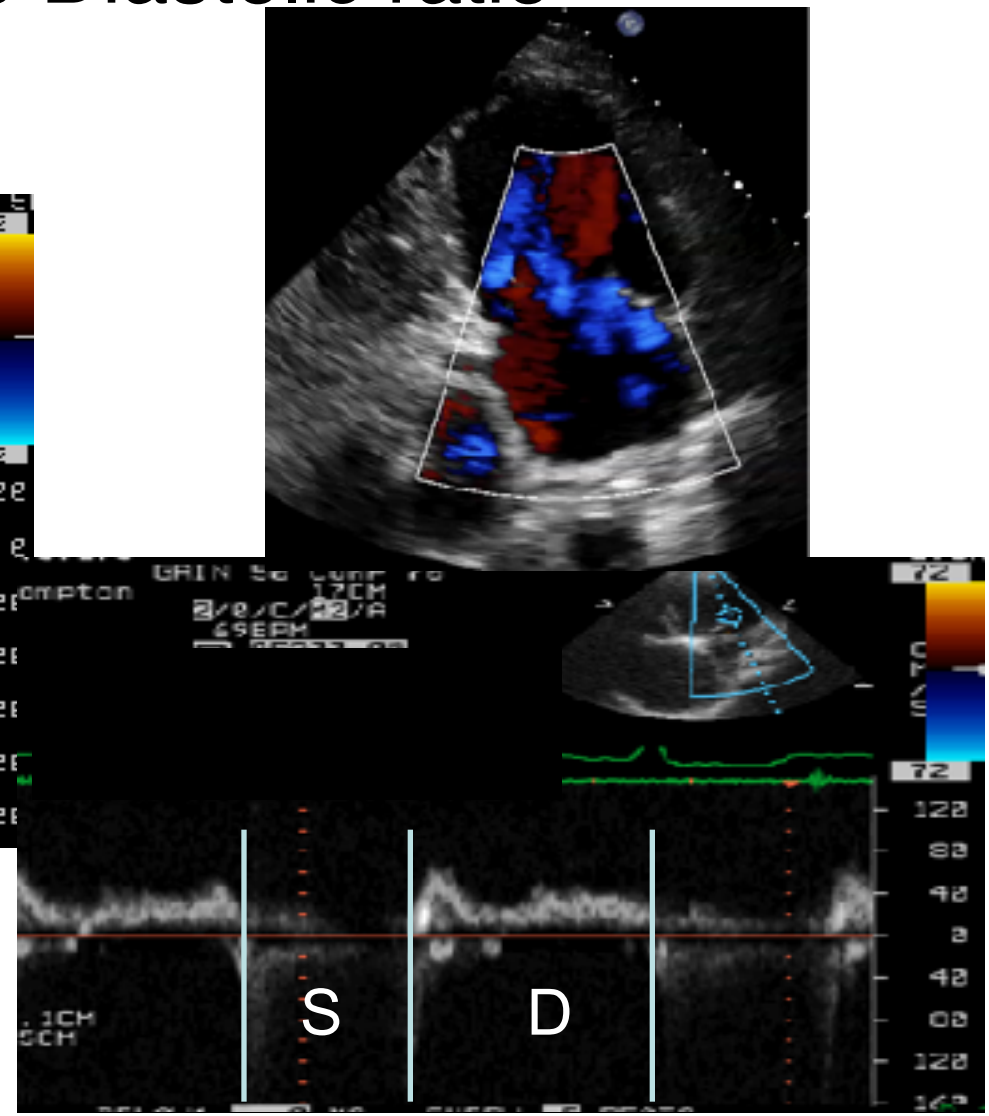
3D Echocardiographic Assessment of Ventricular Volume and Function



Echo Assessment of Ventricular Function
Systolic to Diastolic ratio



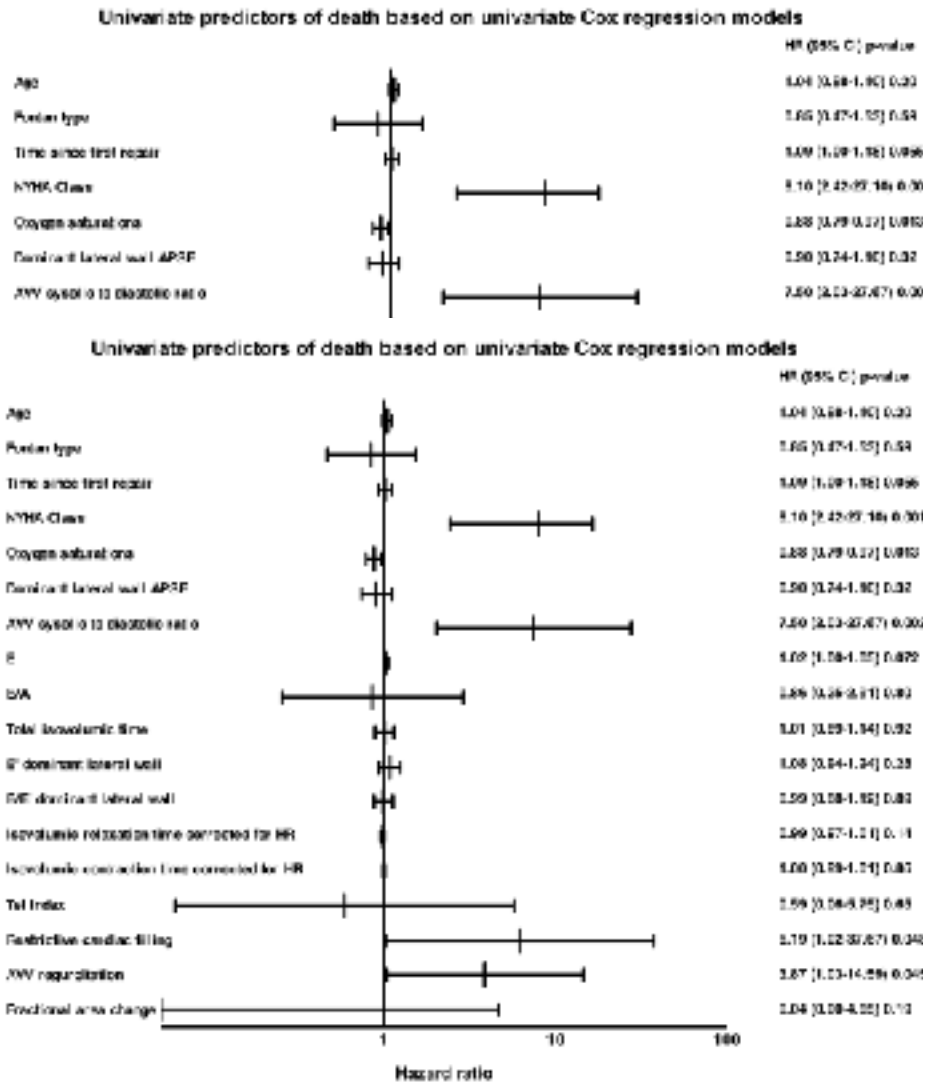
S/D>>1

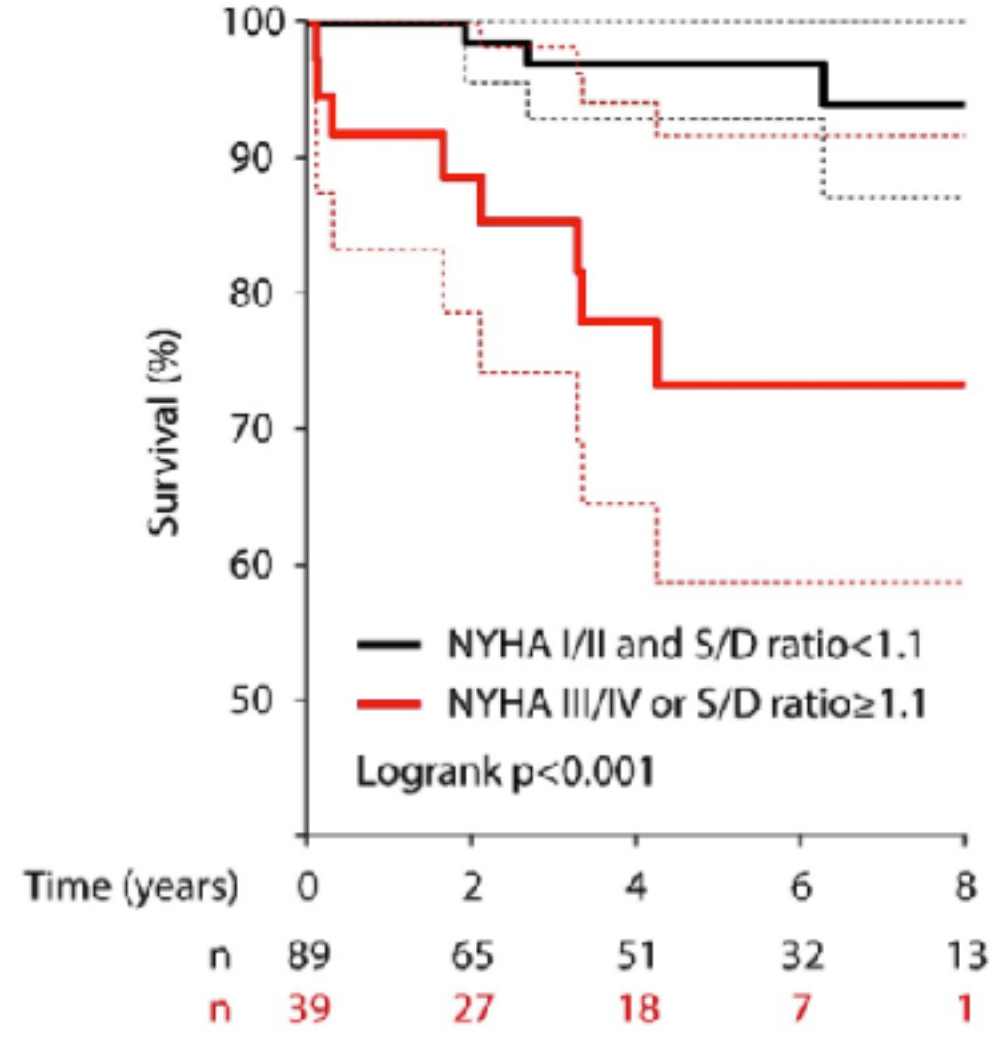


S/D ratio <1

Cordinal R. JACC Imaging 2016

Echocardiographic Predictors of Mortality in adults with Fontan Circulation





Cordinal R. JACC Imaging 2016

Summary

- Echo with its all modalities is a great tool in diagnosis and follow-up of adults with CHD.
- It provides information on cardiac anatomy and physiology.
- 3D Echo offers nonconventional views that are not available with 2D TTE, allow for more accurate assessment of cardiac morphology and function.
- There are still a lot of unknowns in this field.
- Comprehensive assessment depends on complementary imaging modalities.